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ACTA DERMATO-VENEREOLOGICA
VOL. 41. SUPPLEMENTUM 45

CONGENITAL ABSENCE OF VALVES IN THE DEEP VEINS OF THE LEG

A Factor in Venous Insufficiency

PART I

Clinical Observations

by

Anders Lodin

PART II

Roentgenologic Investigations

by

Nils Lindvall and Anders Lodin

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STOCKHOLM 1961

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VOL. 41. SUPPLEMENTUM 45

FROM THE DEPARTMENT OF DERMATOLOGY (HEAD: PROFESSOR SVEN HELLERSTRÖM) AND THE
DEPARTMENT OF DIAGNOSTIC ROENTGENOLOGY (HEAD: PROFESSOR KNUT LINDBLOM)
KAROLINSKA SJUKHUSET, STOCKHOLM

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Translated by Stanley H. Vernon

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HÅKAN OHLSSONS BOKTRYCKERI

Lund 1961

PREFACE

These investigations were conducted at the Departments of Dermatology and of Diagnostic Roentgenology, Karolinska Sjukhuset, Stockholm, and were supported by grants from Edvard Welanders Stiftelse.

To my chief and teacher, Professor Sven Hellerström, I wish to express my deep gratitude for esteemed advice, helpful criticism, and stimulating suggestions during the course of the investigations.

To Assistant Professor Börje Ejrup, who allowed me the use of his laboratory and advised me concerning the investigations of the arterial circulation, I tender my sincere gratitude.

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Finally, I wish to thank Mr Lars Finn for preparation of the illustrations, and Miss Karin Collvin for generous secretarial assistance.

Stockholm, March 1961.

Anders Lodin

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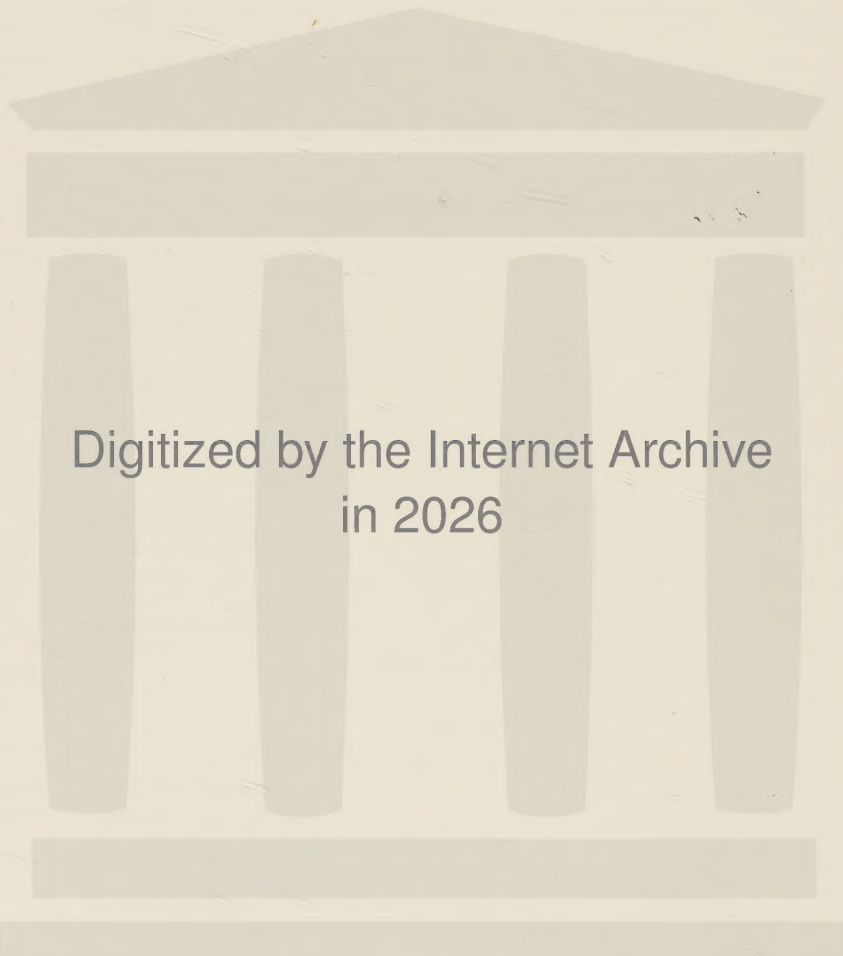
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PART I

CLINICAL OBSERVATIONS



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DEFINITIONS

Vascular lesions, including for instance varicosities associated with edema formation, are of fairly common occurrence in the lower limbs. With protracted standing, the rate of edema formation is increased only slightly above normal, provided the communicating and deep veins have competent valves; but in the case of incompetent valves the rate of edema formation is greatly increased (Holling *et al.*, 1938). If the action of the venous valves is fully eliminated by progressive vascular lesions the tissues will be, in the erect position, more or less constantly subject to an elevated pressure and concomitant chronic edema formation. The complications of this pathologic state include local eczema, cutaneous hemorrhages, pigmentation, sclerosis, and ulcerations. Venous insufficiency is here defined as a chronically impaired venous drainage of this type, attended by various kinds of complications. The term thus includes, whatever the etiology of the insufficiency, those conditions which are variously designated as eczema and/or varicose ulcer, eczema and/or post-thrombotic ulcer, stasis ulcer, the lower leg syndrome, etc.

By "thrombosis" the writer means thrombosis of the deep veins, the term "thrombophlebitis" being reserved for the obstructive and inflammatory processes of the superficial veins, in conformity with current Scandinavian terminology.

INTRODUCTION

The present investigation is based on a study of patients with venous insufficiency associated with *congenital absence of valves in the deep veins of the leg*. The cases were eminently suited for analysis of venous insufficiency, since the symptomatology was not initially obscured by preceding thrombosis, thrombophlebitis or other inflammatory processes in the veins. The aim was to study this anomaly, its incidence and clinical picture (Part I), and its roentgenologic aspects (Part II). Clinical physiologic investigations are now in progress and will be reported at a later date. Their purpose is to study the effects of congenital absence of valves on the local circulation in the legs, as well as the reaction of the general circulation, in order if possible to enhance our understanding of venous insufficiency in the lower leg. In treating here the incidence and social significance, as well as etiologic and pathogenetic aspects of venous insufficiency, the aim is to clarify some of the considerations underlying the investigations which are now proceeding.

Since venous insufficiency in the lower leg is today generally attributed to defective valvular function resulting in an impaired venous return, I include congenital absence of valves as another of its etiologic factors.

A. Incidence and Social Significance of Venous Insufficiency

Pathologic changes in the veins of the leg, leading to venous insufficiency, are very frequently encountered; indeed, many authors consider this morbid condition to be one of the commonest afflictions of civilized peoples. The literature on the subject is copious. Lockhart-Mummery and Smitham (1950) placed the number of patients with leg ulcers in England and Wales at between 100,000 and 200,000. Boyd *et al.* (1952) reported, for England, an incidence of leg ulcers amounting to approximately 5 per mille of the population. Linton (1952) estimated the number of chronic leg ulcer cases in the U. S. A. at 300,000—400,000, while Foote (1954) described varicose ulcers as one of the most disabling diseases in England.

As regards Swedish investigations, Gjöres (1956, 1958) calculated that more than 108,000 persons over the age of 18 (approximately 2.11 per cent of the Swedish population) had suffered from thrombosis of the legs. Of this total, some 102,000 had developed post-thrombotic symptoms and over 31,000 had, or had had, leg ulcers as a result of the earlier thrombosis. On the basis of a six-months series from 1928 Birger (1941) estimated that 300—400 persons annually were granted disability pensions on grounds of varicose veins and leg ulcers. This category represented 1 per cent of the total of approximately 36,000 persons

Table 1. Number of disability pensions initially granted in 1957 and 1958 on grounds of varicose veins and leg ulcers (diagnosis no. 460 according to WHO nomenclature), diabetes (diagnosis no. 260), and rheumatoid arthritis (diagnosis no. 722).

1957																
Cause of disability	Age										Total			Total number of disability pensions granted		%
	—29		30—39		40—49		50—59		60—66					♂	♀	
	♂	♀	♂	♀	♂	♀	♂	♀	♂	♀	♂	♀	♂+♀			
460			1		2	2	7	4	6	14	16	20	36			0.33
260	10	18	20	17	9	11	41	42	52	53	132	141	173	5,851	5,073	1.58
722	10	14	14	28	53	80	109	256	148	218	334	596	930	10,924		8.51

1958																
Cause of disability	Age										Total			Total number of disability pensions granted		%
	—29		30—39		40—49		50—59		60—66					♂	♀	
	♂	♀	♂	♀	♂	♀	♂	♀	♂	♀	♂	♀	♂+♀			
460	1				1	2	3	12	3	19	8	33	41			0.38
260	15	12	21	9	20	5	35	35	52	80	143	141	284	5,979	4,846	2.62
722	5	13	13	27	49	72	110	245	157	249	334	606	940	10,825		8.68

In addition, a total of 45,695 old age pensions were granted — 25,027 of them to males and 20,668 to females — in 1957. The corresponding figures for 1958 were 47,997, 26,332 and 21,665 respectively.

who were granted pensions during that year. By way of comparison, Birger mentioned that the corresponding figure for diabetes was 0.7 per cent.

In order to secure more up-to-date figures, I obtained from the Royal Board of Pensions particulars of disability pensions granted in recent years; they are set forth in table 1. It will be seen that the number of persons who received disability pensions for varicose veins and leg ulcers was 36 in 1957 and 41 in 1958 (0.33 and 0.38 per cent respectively of all disability pensions granted during those years). Important disease categories such as diabetes and rheumatoid arthritis are included in the table for purposes of comparison. The corresponding figures for those diseases were 1.58 and 2.62 per cent, and 8.51 and 8.68 per cent, respectively. As regards leg ulcers the figures are only one-tenth of those reported by Birger 30 years earlier. There may be several explanations of this substantial discrepancy. In the nineteen-twenties, for instance, there was widespread unemployment, as a result of which persons with any physical disability were at a disadvantage in the competition for work. Thus pensions were more likely to be granted before they became due, particularly as the amounts paid at that time were comparatively small. During the nineteen-fifties, on the other hand, full employment prevailed, so that even the partially disabled were able to secure work. The fact that pensions were substantially raised during the same period led, moreover, to more stringent indications for the granting of disability pensions (Jonsson, 1960). The most important factor of all, perhaps, was the legislation of 1948 providing for widows' pensions as from the age of 55, as well as an allowance, as from the age of 60, for the wife of a man drawing any form of pension. As a result of these provisions, a very large category of persons with leg ulcers was eliminated from the disability pension group (Widén, 1960).

Other major factors contributing to the aforementioned discrepancy are the increasingly methodical prophylactic measures against thrombosis and the nowa-

Table 2. *Number of persons in whom varicose veins and leg ulcers (diagnosis no. 460 according to WHO nomenclature), diabetes (diagnosis no. 260), and rheumatoid arthritis (diagnosis no. 722) respectively were diagnosed in 1955.*

Diagnosis no. and sex	Number of cases diagnosed in 1955	Number of sick-pay days until June 1, 1958	Number of sickness periods	Number of compensated sick days before sick benefit became payable
Males	5,200	280,100	6,900	3,200
460 Females	9,000	469,500	11,400	6,200
Total	14,200	749,600	18,300	9,400
Males	2,400	266,000	4,600	4,500
260 Females	1,600	126,200	2,700	3,900
Total	4,000	392,200	7,300	8,400
Males	4,000	683,500	6,900	1,400
722 Females	7,200	1,314,400	11,400	2,400
Total	11,200	1,997,900	18,300	3,800

The investigation from which the tabulated figures are taken is based on a selected series of those members of the sick benefit societies who held *sick-pay insurance* (the series thus includes only occasional members aged 67 or over). The selection comprises those born on the 15th of a month, and in the above table the figures calculated on that basis have been multiplied by thirty.

days routine treatment of thrombosis with anticoagulant drugs. In Sweden, such therapy was initiated with heparin, the original coagulant, purified by Jorpes (1935). When thrombosis does develop, it invariably appears to have its origin in the deep venous channels of the lower leg — according to Foote (1954), usually in the posterior or anterior tibial vein — and spread of the process to the thigh generally occurs after a short time (Bauer, 1942, and others). In Bauer's opinion (1957) this upward spread of the thrombosis through the femoral vein takes place in at least four of five untreated cases. The possibility of preventing it by early anticoagulant therapy, and hence of precluding post-thrombotic sequelae has been emphasized by numerous authors (Bauer, 1940, 1941, 1951, 1955 a; Jorpes, 1946, 1954; Brennhovd, 1956; Gjöres, 1956, 1958; Gilje 1958; and many others).

Yet another factor of significance in this connection may be more widespread treatment of post-thrombotic conditions by recent methods such as e.g. the Bisgaard bandage (1939), the Birger bandage (1947), the adhesive tape, sponge rubber and elastic bandage technique of Gilje (1949, 1958), and the Rastgeldi boot (1957).

The significance of leg ulcers as a cause of incapacity for work is illustrated by table 2, based on figures obtained from the National Insurance Office. The estimated total of 14,200 persons who developed leg ulcers in 1955 had, over a three-year period, sick leave totalling in round figures 750,000 days, or almost two months per individual. The corresponding figures for diabetes and rheumatoid arthritis were three months and six months respectively. Sweden has a population of about seven and a half million. Roughly a quarter of a million

Table 3. *Number of patients with diagnosis no. 460 (varicose veins and leg ulcers) according to WHO nomenclature, who were hospitalized at the Dermatology Department of Karolinska Sjukhuset during the period 1955—1959, and duration of hospitalization.*

Years	Males	Females	Total	Total admissions	%	Number of hospitalization days			Average hospitalization time in days		
						Males	Females	Total	Males	Females	Total
1955	24	102	126	1,694	7.4	677	4,229	5,606	28.2	41.5	44.5
1956	27	83	110	1,591	6.9	1,543	4,391	5,934	57.1	52.9	53.9
1957	24	73	97	1,362	7.1	828	3,008	3,836	34.5	41.2	39.5
1958	32	108	140	1,665	8.4	1,231	4,013	5,244	38.5	37.2	37.5
1959	36	83	119	1,410	8.4	1,557	3,535	5,092	43.2	42.6	42.8

working days are lost each year because of venous insufficiency in the lower leg.

It will be seen from table 3 that no less than about 7 per cent of all patients admitted to the Dermatology Department of Karolinska Sjukhuset have suffered from leg ulcers due to venous insufficiency. The average hospitalization time has varied between one and two months. These figures demonstrate the social significance of the disease.

Jorpes (1946), it may be pointed out, demonstrated that in Sweden disability due to post-thrombotic conditions was more common than disability resulting from automobile accidents.

The foregoing data make it quite evident that venous insufficiency is of major social significance, particularly as it afflicts persons of all vocations and social groups, frequently at the most productive ages.

B. Etiologic and Pathogenetic Considerations

A very large number of investigators have, over the course of the years, studied problems associated with venous insufficiency in the lower leg. Its ultimate complication, ulcer of the leg, is referred to in the earliest medical literature. Even Hippocrates, according to Anning (1954), observed the relationship between varicose veins and leg ulcers. In the course of time, numerous theories have been propounded to account for their causes, such as pathologic composition of the blood, abnormal blood supply, inhibition of the venous drainage, and abnormal pressures in the peripheral vascular system.

An etiologic relationship between venous stasis and leg ulcers was surmised as early as the seventeenth century. In 1676, for instance, Wiseman wrote of venous thrombosis, the dilatation of veins, and the effects of those conditions on the valves. He appears to have been the first to realize that leg ulcers could be the direct result of a circulatory defect, and he employed the term "varicous ulcer". Most authors during recent years have apparently subscribed to the view that the ulcers are due to stasis. Indeed, the term "stasis ulcer" is frequently used in American literature.

Although the ulceration was formerly attributed primarily to the varicose veins (varicose ulcers of the leg), the last few decades have brought a number

Table 4. *Percentual distribution of post-thrombotic, varicose and other types of leg ulcers.*

Author	Number of leg-ulcer cases	Post-thrombotic %	Varicose %	Other types %
Birger 1941	432	33.1	45.4	21.5
Bauer 1942	38	87	13	0
Nilzén 1945	111	56*		
Birger 1947	869	40	45	15
Gilje 1949	626	33.87	52.24	13.89
Stürup 1950	335	38.8	44.2	17
Anning 1954	1,026	74.9	10.7	14.4

* including 15 per cent uncertain.

of works which show that the varicose syndrome is based to at least the same extent upon *previous venous thrombosis* followed by recanalization with formation of an irregular, avalvular venous channel.

According to Bauer (1951) recanalization "seems to be a rule with very few, if any, exceptions".

Scandinavian researchers have expended considerable labor on studies in this field. Data from a few of the multitudinous publications are listed in table 4.

There is another group of leg ulcers in which no insufficiency is demonstrable either in the superficial or in the deep venous trunks, whether anamnestically or by means of conventional clinical or roentgenologic procedures. The British author Cockett (1953) communicated a study of such cases in which he demonstrated the pathogenetic significance of post-thrombotic lesions of the communicating veins between the superficial and deep venous systems in the distal part of the lower leg (Cockett perforators).

In recent times another condition has been suggested — though not universally accepted (Dohn, 1958) — as a cause of venous insufficiency; namely, non-post-thrombotic deep incompetence (Martorell, 1946, 1951; Bauer, 1948, 1950, 1951, 1955 b, 1957, 1958 and 1960; Lockhart-Mummery and Smitham, 1950; Dohn, 1956; and others). This pathologic state is characterized by the presence of incompetent valves in the deep veins. Since it is largely through the investigations of Bauer that the manifold problems associated with this condition are approaching solution, his concept of the syndrome, for which he proposes the term *pan-phlebo-sis*, will be adhered to here. Bauer found that almost one-half of the cases with post-thrombotic symptoms in his extensive series had not suffered from deep venous thrombosis but rather were victims of pan-phlebo-sis. According to Bauer, such cases show an hereditary predisposition to varicose veins. Females predominate, outnumbering males by approximately two to one. There is nothing to indicate that pregnancy has any bearing on the condition. Clinical examinations yield findings almost identical with those in post-thrombotic cases. The phlebograms, however, differ markedly, the femoral vein shadow being of ample and uniform width and showing soft contours. Numerous

valve formations are generally observed. At operation the walls of the femoral and popliteal veins are found to be grossly normal or slightly thickened, and the lumen in no way affected. Microscopic examination reveals thickening of the intima with generalized hyaline fibrosis and splitting of the elastic elements. The media too shows marked fibrosis, causing splitting of the elastic elements and atrophy of the muscular layer.

In Bauer's opinion these changes are due to an apparently specific degenerative process in the vein wall, resulting in loss of elasticity. In this way the involved segment of vein may be abnormally distended by its contents; in consequence the valve leaflets are no longer able to meet in apposition but become incompetent. This soon results in further distension of the vein, and a vicious circle arises. Bauer has a series of more than 2,000 microscopic analyses of excised pathologic veins, and the findings are homogeneous irrespective of the venous groups to which the specimens are assignable. He summarizes the syndrome as follows:

1. It is chronic.
2. It affects in greater or less degree the majority of veins in the leg, both deep and superficial.
3. It is characterized by an insidious, sometimes more rapidly progressive, degenerative process in the vein wall, involving in particular the elastic elements.
4. Its etiology is unknown except for the fact that hormonal influences can be traced.

CHAPTER 2

NORMAL ANATOMY OF THE VENOUS VALVES IN THE LEG

Recent years have brought numerous detailed reports on the historical aspects of varicose veins, venous thrombosis, and ulceration of the leg (*vide e. g.* Raivio, 1948; Gilje, 1949; Anning, 1954; Foote, 1954). According to the historical studies of Steudel (1955), a number of European workers discovered the venous valves more or less contemporaneously in the mid-sixteenth century.

Modern textbooks of anatomy contain little or no mention of the normal incidence of venous valves in the lower extremities. A brief outline will accordingly be presented here.

All workers who have reported studies of venous valves seem to agree that major individual variations exist. Usually the veins of the two limbs are equally valved, though considerable variability both as to number and localization may be found, not only in different persons but in the two extremities of a single subject. As a general rule, however, the most common localization of valves in a major vein is immediately below its junction with a branch, and in a branch, immediately before its termination.

According to Gray (1946), each valve is formed by a reduplication of the inner coat, strengthened by connective tissue and elastic fibers, and is covered on both surfaces with endothelium. Most commonly, two such semilunar valves are found opposite one another; occasionally there are three, and sometimes only one. — When in this paper, as in the works of other authors, reference is made to “valves” the term generally connotes a pair of valves.

A. Deep Veins above the Knee

1. The *inferior vena cava* is devoid of valves.

There appears to be complete unanimity of opinion that this vein is invariably avalvular (M'Dowel, 1849; Wahlgren, 1851; Trendelenburg, 1880; Bardeleben, 1880; Friedreich, 1882; Poirier, 1896; Hesse and Schaack, 1909; Testut, 1921; Tandler, 1926; Rauber and Kopsch, 1941; Eger and Casper, 1943; Eger and Wagner, 1949; Basmajian, 1952; Foote, 1954; Hamilton, 1956; and others).

2. The *common iliac vein* is usually avalvular.

Most authors consider this vein to be avalvular. Several researchers, however, have demonstrated valves in it. Friedreich (1882), for instance, found single valves in five of 370 veins (1.4 per cent) — symmetrically bilateral in one case, on the right side alone in two, and on the left side alone in one. Waldeyer (1899)

reported a figure of 1—2 per cent for valves in this vein. McMurrich (1906) arrived at similar results, finding valves in two of 60 veins investigated. According to Warwick (1930, 1931) the common iliac vein occasionally has one valve, which for the most part is incompetent. Patten (1943) points out that though valves may occur, they are usually absent. DiDio (1949) in his meticulous studies of 160 cadavers, found valves in the common iliac vein in 5.6 ± 1.81 per cent of the cases. Most of these valves were incompetent. Basmajian (1952), without having found a single valve himself, remarked that valves in the common iliac vein "are not as rare as it was believed formerly, but few, if any, are competent". This view seems to have been based entirely on the above-mentioned investigation conducted by DiDio.

3. *The external iliac vein* contains one valve in approximately 25 per cent.

Friedreich (1882), on studying the external iliac veins in 185 cadavers of both sexes and varying age, found that valves, if present, were often incompletely developed. In 41 cases (just over 22 per cent) he observed a valve situated bilaterally, usually at symmetrical sites, and in 48 cases (26 per cent) a unilateral valve — on the right in 36 and on the left in twelve. Thus valves were found in 130 (35 per cent) of the 370 veins studied. Waldeyer (1899) in pointing out the presence of valves in the common iliac vein in 1—2 per cent, added that "*öfter findet man ein Klappenpaar am Beginne der Venae iliacaе externaе*" (more often a pair of valves is found at the start of the external iliacs). McMurrich (1906) stated that in isolated instances two valves might occur in this vein, though he found it to be avalvular in more than one half of 60 specimens examined. In many cases the valves were clearly inefficient. Quain (1894), like Warwick (1930) and Gray (1946), believes that the external iliac vein is "frequently provided with one valve". Eger and Casper (1943) found a single valve in just under one-half of the 38 cadavers which they studied. Eger and Wagner (1949) reported a 24 per cent incidence of solitary valves in 200 veins from subjects of varying ages, and Powell and Lynn (1951) found a 33 per cent incidence in 54 limbs. The localization was invariably just distal to the junction with the internal iliac vein. Basmajian (1952) was able to demonstrate a single valve in 16 of 72 veins studied (22.2 per cent). Twelve of the valves were located within 3 cm of the junction with the internal iliac, and the remaining four within a distance of 3 cm from the inguinal ligament; 13 were found on the right and three on the left side. Summarizing, Basmajian stated that "judging from the accumulated data of four independent studies involving many hundreds of veins, one in four external iliac veins (24 per cent) contains a valve. More than two-thirds of these are competent".

It should, however, be made clear that several authors, particularly the early ones, consider the external iliac vein to be avalvular. Hesse and Schaack (1909), for instance, found no valve in any one of 200 veins examined.

4. *The femoral vein* contains an average of four valves; the number may vary between one and nine.

Wahlgren (1851) states that the valves in this vein number two, while according to Houzé (1854) they are three in number. Bardeleben (1880) in his investigations found that the number varied between one and five, and gave

an average of four or five. M'Dowel (1849) who, according to Bardeleben, was the first to report the number of valves in a specific vein, said that in the femoral vein the number varied between three and five.

Friedreich (1882) reported an incidence of 81 per cent for valves in the upper five centimeters of the femoral vein. In his opinion the occurrence of competent valves in the proximal part of that vein may be considered the rule. He claimed to have found in each of the 370 femoral veins examined, a valve immediately above the junction with the deep femoral, a localization which he alone seems to have observed. According to Quain (1894) the femoral vein is provided with three to five valves. Poirier (1896) notes the almost constant occurrence of an ostial valve, in addition to one to five others occurring in the remaining part of the femoral vein. Hesse and Schaack (1909) usually found one valve (72 per cent) and occasionally even two valves (5 per cent) in the femoral vein above its junction with the long saphenous vein. In 23 per cent, however, that part of the femoral vein was avalvular. In each of the 100 cadavers which they studied, valves were found distal to the junction with the long saphenous vein: two valves in 73 cases, three or more in 18, and a solitary valve in nine cases. The average distance between the valves was 3—4 cm. The first valve in this sector of the femoral vein was found, on the average, 3—5 cm below the junction of the long saphenous vein.

In the opinion of Patten (1943) the femoral vein usually contains about five valves. According to Raivio (1948) the number varies between four and six, whereas Powell and Lynn (1951) mention a range of one to four. The latter authors found one valve in 10 per cent, two in 33 per cent, and three or more in 57 per cent. Basmajian (1952) observed an average of three valves between the inguinal ligament and the adductor canal. In one of his cases (1.3 per cent) valves were quite absent. He occasionally noted as many as six valves (in 6.6 per cent) or as few as one (in 7.9 per cent). Most of the veins contained two valves (23.8 per cent), three valves (33 per cent) or four valves (23.8 per cent). According to Foote (1954) the number of valves in the femoral vein is one to five, whereas Dodd and Cockett (1956) report a range of two to nine and an average of five.

Quoted above is a report by Friedreich (1882), who states that the femoral vein usually contains a valve just *above* the deep tributary. The most common site of a valve in the femoral vein is, however, just *distal* to the mouth of the deep tributary, at which location Powell and Lynn (1951) found a valve in over 93 per cent and Basmajian (1952) in about 90 per cent of cases studied. Most authors are in agreement on this point (e. g. Kampmeier and Birch, 1926; Warwick, 1930; Eger and Wagner, 1949; Boyd *et al.*, 1952; Foote, 1954; Dodd and Cockett, 1956). The next most common site is at or just distal to the inguinal ligament, where the majority of femoral veins have a valve (Houzé, 1854; Poirier, 1896; Hesse and Schaack, 1909; Warwick, 1930; Powell and Lynn, 1951; Basmajian, 1952; Foote, 1954; Dodd and Cockett, 1956, and others). Gullmo (1957), among others, believes that a valve, frequently termed the "sub-Poupartian valve", is normally present in the femoral vein 0.5—1 cm distal to the inguinal ligament.

There have also been a number of roentgenologic studies of valves in the veins of the lower extremities. Greitz (1955), for instance, using ascending

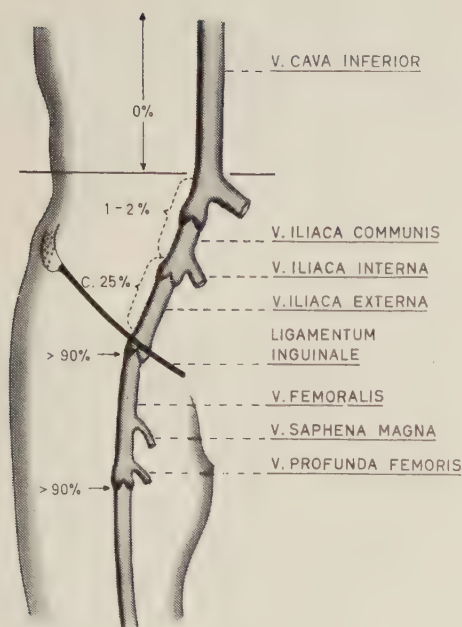


Fig. 1

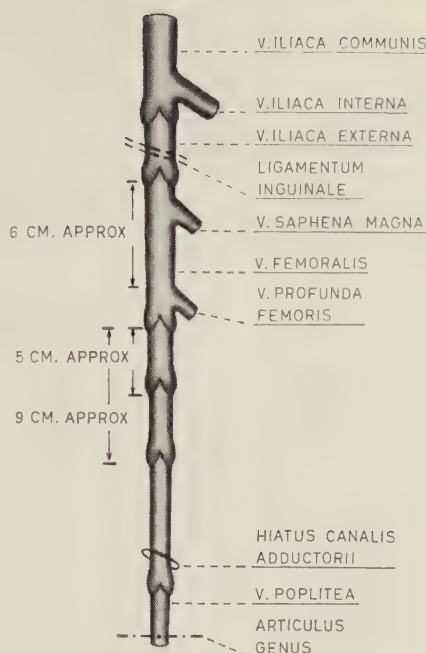


Fig. 2

Fig. 1. Incidence of valves in the inferior vena cava, common and external iliac veins, and at two special sites in the femoral vein. Modified after Basmajian (1952).

Fig. 2. Interrelations of valves in the deep veins of the thigh. Modified after Powell and Lynn (1951).

phlebography in 30 normal extremities, found one to five valves in the femoral vein, most of them at proximal sites. Mathiesen (1958) reports having observed, in tilt phlebography of 27 normal legs, between two and seven valves, the average being 4.2 in each thigh. These findings accord closely with the results of the anatomic dissections outlined in the foregoing.

As regards the *deep femoral vein* it may be mentioned that Bardeleben (1880) reports a figure of four valves, one of them invariably situated just below the junction with the femoral vein. Poirier (1896), Warwick (1930) and others concur with this finding.

The most common sites of valves in the above-mentioned veins are shown schematically in figures 1 and 2.

5. The *popliteal vein* contains an average of two valves, the range being from one to five.

M'Dowel (1849) found four to five valves in this vein, whereas Wahlgren (1851) had a figure of two. Quain (1894) and Patten (1943) give the number as two to three. Gray (1946) states that "there are usually four valves in the popliteal vein". Powell and Lynn (1951) in studies limited to the proximal third of the popliteal vein, found in that sector a valve just distal to the adductor ring in 96 per cent of the cases. A similar finding had been reported by Warwick

(1930), who succinctly observes that "the beginning of the popliteal vein is valved". Foote (1954) notes that the number of valves throughout the popliteal vein varies between one and four; Dodd and Cockett (1956) as well as May and Nissl (1959) report a range of one to three, most cases having two. Similar figures have also been recorded by early investigators such as Houzé (1854), Poirier (1896), Testut (1921) and others. Bardeleben (1880), however, observed as a rule only one valve in the popliteal vein. According to the consensus, one valve invariably occurs just distal to the junction of the short saphenous vein.

B. Deep Veins of the Lower Leg

Numerous authors report that the deep paired veins of the lower leg are those most plentifully supplied with valves. The individual variations, however, are substantial. M'Dowel (1849) is content to observe that both the deep and the superficial venous systems of the leg "are likely provided with valves, which are most numerous in the veins of the former class"; while Wahlgren (1851) thinks that the deep veins of the lower leg have valves for almost every half inch of their length. An early edition of Quain's *Anatomy* (1866) contains the statement that in the lower leg "the valves of the deep veins are very numerous — ten or twelve being sometimes found between the heel and the knee". Bardeleben (1880) notes quite briefly that these veins are always well provided with valves, and Dodd and Cockett (1956) consider them to be "profusely valved, the valves being situated about every inch or so". Rauber and Kopsch (1941) observe in their textbook only that valves occur abundantly in veins of the leg, "*doch sind sie bei den tiefen zahlreicher*" (though their number is greater in the deep veins). Pattern (1943) and Gray (1946) believe that all deep veins of the lower leg contain numerous valves. Dissections made by Rémy (1901) showed about 18 valves in each pair. Greitz (1955) in phlebography of 30 normal legs invariably observed valves in the deep veins of the lower leg — "ten or slightly more in each paired vein".

1. The *posterior tibial veins* contain on the average approximately ten valves each, the number ranging from seven to twenty.

Quain (1866) reports ten to twelve valves in these veins, whereas Bardeleben (1880) found a range of seven to sixteen and an average of ten. Poirier (1896) says the number varies from eight to twenty. Raivio (1948) gives a figure of seven to nine, which in his opinion is relatively low as compared with other published figures. His tabulation of the figures recorded by different authors indicates approximately the same variation, the extremes being within the range reported by Poirier.

2. The *anterior tibial veins* contain an average of about ten valves each, the number varying from nine to twelve.

Houzé (1854) reports a figure of eleven, Quain (1866) a figure of ten to twelve, and Bardeleben (1880) a range of nine to twelve, with eleven as the average. According to Poirier (1896) there are at least eleven valves in these veins. Rémy (1901) puts the figure at nine, and Greitz (1955) at ten or slightly more.

3. The *peroneal veins* each contain, on the average, approximately ten valves, the range being from six to twelve.

Houzé (1854) notes that in these veins the number of valves is at least eight; Quain (1866) has ten to twelve, but Bardeleben (1880) records only six. Poirier (1896) reports eight to ten, Rémy (1901) nine, and Greitz (1955) approximately ten or slightly more.

C. Superficial Veins of the Leg

1. The *long saphenous vein* contains an average of ten valves, the range being from seven to twenty.

Raivio (1948) states in his doctoral dissertation that the number of valves in the long saphenous vein varies between one and seven (average 3.5) in the thigh, between two and six (average 4.0) in the lower leg, and between one and four (average 2.9) in the foot; the range for the entire vein being seven to fifteen and the average 10.4. Raivio's survey of the data presented by a large number of authors shows close agreement with his own findings, and subsequently published investigations including those of Foote (1954) and Dodd and Cockett (1956) are likewise in accord. Worthy of note is that Quain (1894) records as high a number as seven to twenty and Gray (1946) ten to twenty.

In a Japanese series comprising 113 cadavers (89 males and 24 females) ranging in age from 15 to 91 years, Ogo (1933) found in each case valves at the opening of the long saphenous into the femoral vein. The above-mentioned reports concerning the occurrence of a valve in a branch below its junction with a larger vein, may well apply to the long saphenous vein as well. Views consistent with Ogo's are expressed by the majority of authors (Wahlgren, 1851; Houzé, 1854; Braune, 1871; Trendelenburg, 1880; Bardeleben, 1880; Klotz, 1887; Poirier, 1896; Hesse and Schaack, 1909; Kampmeier and Birch, 1926; Rouvière, 1927; Warwick, 1930; Raivio, 1948).

2. The *short saphenous vein* is provided with an average of eight valves, the range being from five to fifteen.

According to Raivio (1948), the number of valves in this vein varies from five to thirteen and averages 8.2. The range in the foot region is three to seven and the mean number 4.3. The corresponding figures for the lower leg are two to six and average 3.9. Here too, Raivio's values closely accord with the data which he cited from the works of a number of different investigators. These data in turn are in close agreement with those given in subsequent papers (e. g., Foote, 1954). It should be pointed out, however, that several authors report somewhat greater numbers. Poirier (1896), for instance, gives a figure of nine to ten for the lower leg, and of two to three for the foot. The corresponding figures recorded by Dodd and Cockett (1956) are six to twelve and two to three respectively. In the opinion of Testut (1921) there are eight to fifteen in the entire vein. Just as the long saphenous vein has a valve below its junction with the femoral vein, so the short saphenous has, as a rule, a valve just below its junction with the popliteal vein.

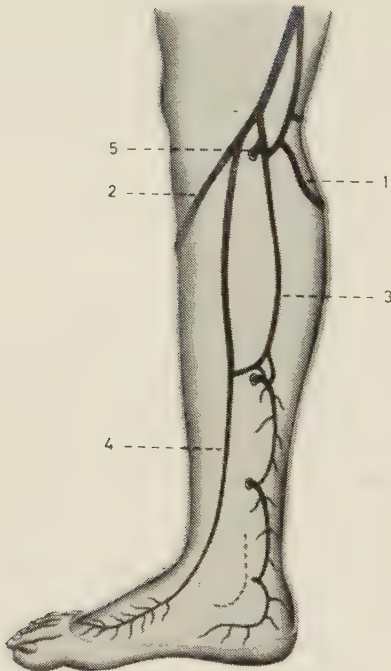


Fig. 3

Fig. 3. The long saphenous and medial perforating veins.

1. The connection with the short saphenous vein.
2. A tributary from the anterior surface of the leg.
3. The posterior arch tributary which also links the three medial perforating veins.
4. The long saphenous vein.
5. A constant direct communicating vein.

After Dodd and Cockett: *The Pathology and Surgery of the Veins of the Lower Limb*.
E. & S. Livingstone, Ltd., Edinburgh, 1956.

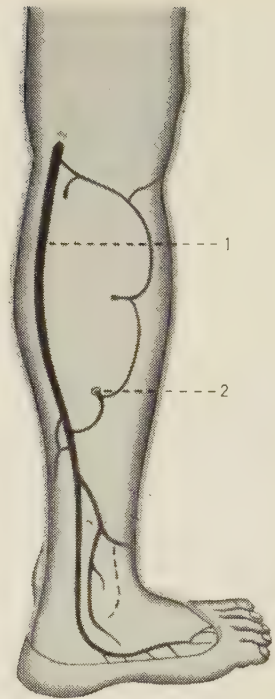


Fig. 4

Fig. 4. The usual arrangement of the lateral perforating veins. There is usually one large constant perforating vein (2) in the position shown, 4 to 7 inches above the tip of the lateral malleolus at the outer border of the triceps surae tendon. It usually connects with the short saphenous vein (1). After Dodd and Cockett: *The Pathology and Surgery of the Veins of the Lower Limb*. E. & S. Livingstone, Ltd., Edinburgh, 1956.

D. Communicating Veins

In his dissertation on the communicating veins of the leg Raivio (1948), like Linton (1938) before him, makes a distinction between tibial, fibular and dorsal communicating veins. Höjensgård (1951) suggests that they should be termed medial, lateral and dorsal communicating veins, a proposal which Greitz (1955) finds highly practical from the roentgenologic standpoint. According to Raivio, the number of these communicating vessels between the superficial and the deep veins of the thigh and the lower leg is approximately eighty, most of them situated medially and laterally in the calf. The number of valves, in his view, varies between one and five, but is commonly two. All valves are directed

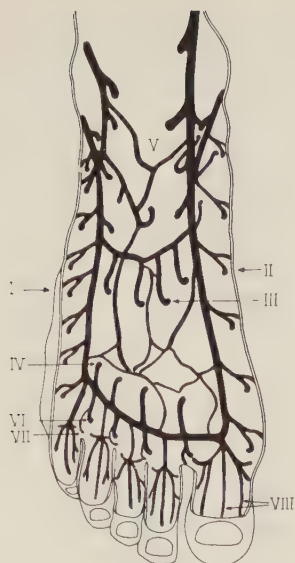


Fig. 5. Communicating veins of the foot.

I.	Vv. communicantes marginales fibulares	(8)
II.	" " tibiales	(6)
III.	" " metatarsales proximales	(4)
IV.	" " " distales	(4)
V.	" " dorsales pedis	(13)
VI.	" " intercapitulares profundi	(4)

After Raivio (1949).

toward the deep veins, thus preventing reflux towards the skin. As regards the anatomy of these veins the reader is referred to the detailed account presented by Raivio. Suffice it here to outline certain data of importance with respect to venous insufficiency; for the rest, reference should be made to Linton and Raivio as well as e. g., Sherman (1949), Dow (1951), Foote (1954), Dodd and Cockett (1956) and May and Nissl (1959).

Of the communicating veins, the direct ones which unite the saphenous trunks with a deep vein are clinically the most important. The indirect ones communicate first with a muscle vein, and thence a further vessel connects with the main deep trunk.

The most important of the direct perforating veins number, in the thigh, up to six and are referable to the tibial group. The principal one is considered to be that situated at the level of the adductor canal.

In the knee region there are about ten of these veins, connecting the saphenous veins with the popliteal vein. An important perforator of almost invariable occurrence is that which passes from the long saphenous vein along the posterior margin of the tibia to the posterior tibial vein (figure 3: 5).

On the medial aspect of the lower leg there are up to ten communicating veins between the long saphenous and the posterior (or anterior) tibial vein; on the anterior aspect, approximately the same number between the saphenous veins and the anterior tibial; and on the lateral aspect up to eight between the short saphenous and the fibular veins.

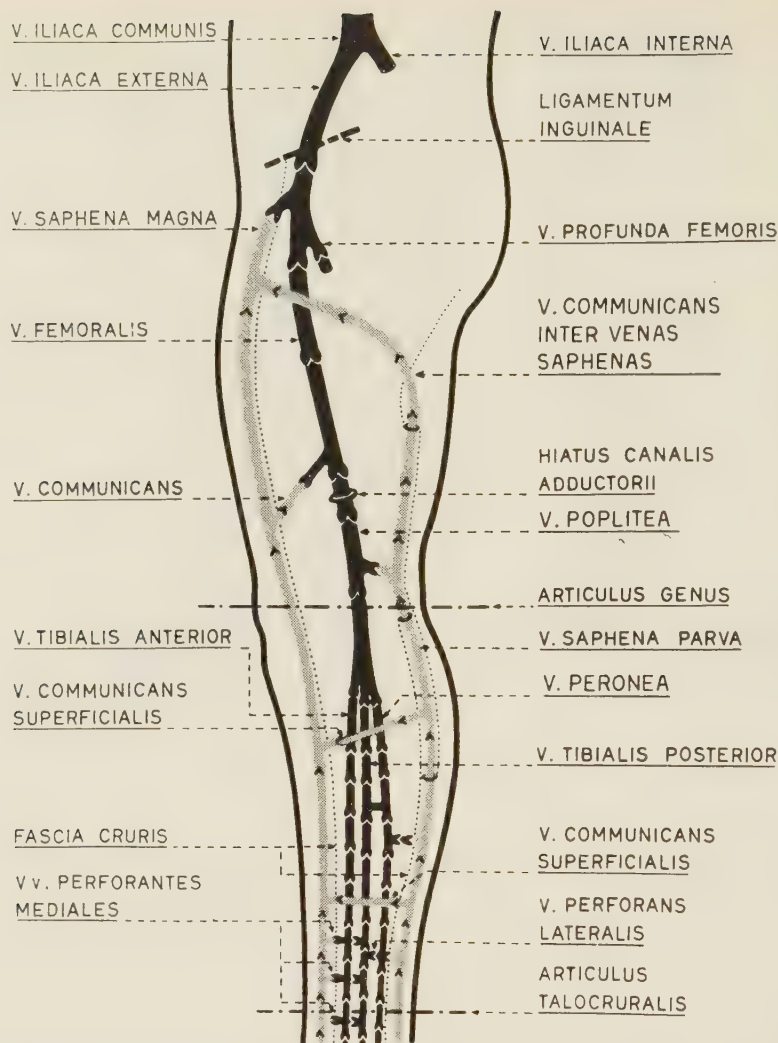


Fig. 6. A diagrammatic rendering of the veins of the lower limb. Modified after Dodd and Cockett: *The Pathology and Surgery of the Veins of the Lower Limb*. E. & S. Livingstone, Ltd., Edinburgh, 1956.

The chief perforators of the lower leg are known as the internal and external ankle perforating veins, or "Cockett perforators". The internal ones receive blood from the lower and inner third of the leg and terminate in the posterior tibial vein. Not until recent years has attention been drawn to the causal significance of valvular incompetence in these perforators with respect to edema and leg ulcers. A schematic representation of these vessels will be found in figures 3 and 4.

The number of communicating veins in the foot between the superficial and the deep venous system is very substantial — according to Raivio, forty or so (figure 5).

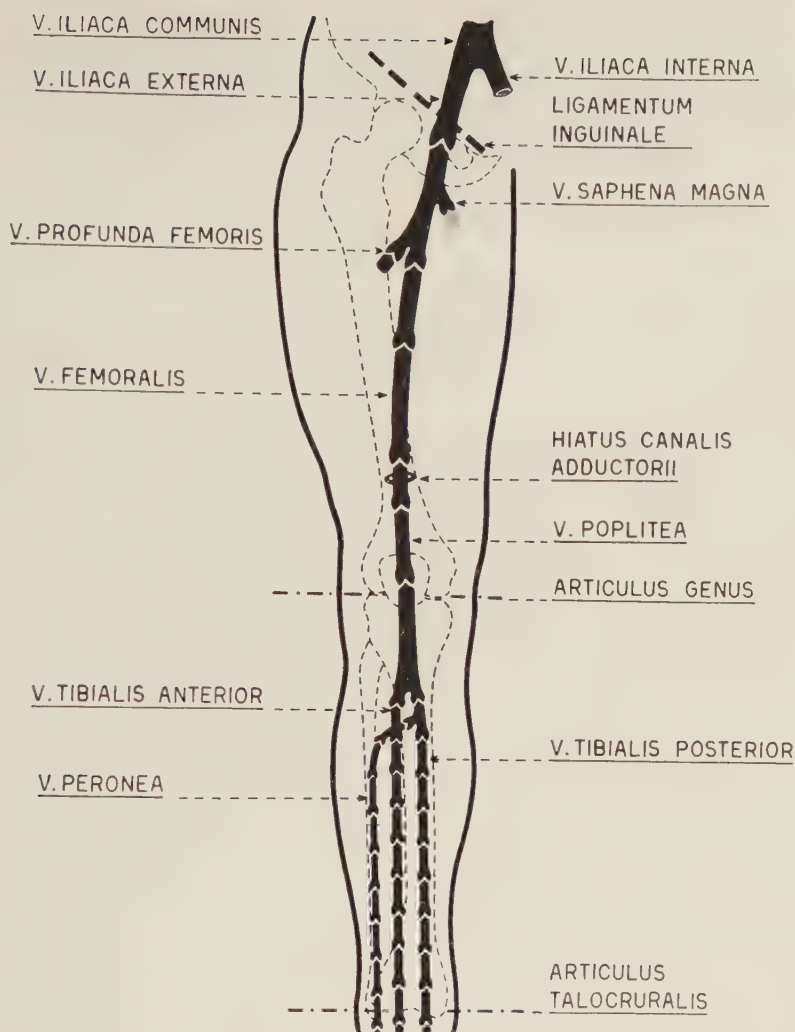


Fig. 7. A diagrammatic rendering of the deep veins of the lower limb.

There is no significant difference in the number of venous valves in males and females (Kosinski, 1926; Raivio, 1948; Powell and Lynn, 1951, and others), although some authors think otherwise; Ogo (1933), for instance, states that venous valves are generally more numerous in females than in males.

The claim, notably by early investigators (Bardeleben, 1880; Braune, 1884, Klotz, 1887; Poirier, 1896; Testut, 1921, and others) to the effect that the number of valves is progressively reduced with mounting age has been strongly refuted by more recent authors (e. g. Kosinski, 1926; Kampmeier and Birch, 1926; Berntsen, 1927; Ogo, 1933). Powell and Lynn (1951), in common with many other investigators, found in their anatomic dissections a varying number of valves in the deep veins of the fetus and the adult, but no significant dif-

ference in number with respect to increasing age. A similar observation was made by e. g. Dodd and Cockett (1956). The above-mentioned authors, on the other hand, express opinions which seem to be consonant with Kosinski's observation that "with age some valves are slightly degenerated and the walls of the veins are thickened".

Summary

The <i>inferior vena cava</i>	is avalvular.
The <i>common iliac vein</i>	is, as a rule, avalvular.
The <i>external iliac vein</i>	is avalvular in 75 per cent.
The <i>femoral vein</i>	has an average of 4 valves, the range being 1—9.
The <i>popliteal vein</i>	" " " " 2 " " " " 1—5.
Each <i>posterior tibial vein</i>	has an average of approximately 10 valves, the range being 7—20.
Each <i>anterior tibial vein</i>	has an average of approximately 10 valves, the range being 9—12.
Each <i>peroneal vein</i>	has an average of approximately 10 valves, the range being 6—12.
The <i>long saphenous vein</i>	has an average of 10 valves, the range being 7—20.
The <i>short saphenous vein</i>	" " " " 8 " " " " 5—15.
The <i>communicating veins</i> ,	approximately 80 in number and situated largely in the calf, have, as a rule, two valves.

On the basis of the foregoing survey the normal incidence of valves in the veins of the legs may be schematically represented as shown in figures 6 and 7.

CHAPTER 3

PREVIOUS INVESTIGATIONS INTO CONGENITAL ABSENCE OF VALVES IN THE DEEP VEINS OF THE LEG

It is evident from the foregoing review that numerous individual variations may be found in respect to the extent and number of the venous valves. Wahlgren (1851), for instance, advances the view that these variations are related to the varying development of the venous system: In physically strong subjects, he points out, one finds valves in veins which in physically weak persons are not valved; sometimes, moreover, veins which normally have valves are found to be avalvular; and indeed on rare occasions this deficiency has been found to involve the entire venous system. Wahlgren was referring to an investigation conducted by Hodgkin and Cooper, who in 1836 had described two acephalic monsters who were totally devoid of venous valves.

The idea has also been advanced of partial or total congenital absence of valves in the superficial and/or deep veins of the legs as a cause of venous insufficiency. Kraemer (1898) on the basis of experience from 27 surgically treated cases of varicose veins, concludes that a "*congenitale Anlage in einer mangelhaften Klappenbildung*" (congenital predisposition to defective valve formation) in the long saphenous vein may be an etiologic factor in varices. In seven cases he had studied the veins following excision. Five of these seven were entirely devoid of valves and two had a single valve. Kraemer observed, however, that "*hierbei berücksichtigte ich nur den Verlauf am Oberschenkel bei bloss geringfügiger Erweiterung; die vorgeschritteneren Fälle, wie meist am Unterschenkel, waren natürlich nicht zu gebrauchen*". (I took only the course in the thigh into account when there was insignificant dilatation; the far advanced cases, usually involving the lower leg, were naturally unserviceable.) It was earlier pointed out that of the 100 cadavers studied by Hesse and Schaack (1909), 23 had no valves proximal to the junction of the long saphenous with the femoral vein. Those authors consider this deficiency to be congenital and attach major importance to it in the etiology of varices.

Warwick (1931) writes in his textbook that the perforating veins in connection with the long saphenous vein, and likewise certain indirect perforating veins of the calf (the muscle veins), may be devoid of valves. In such a case the superficial veins in the leg are not protected against the rapid rises of intravenous pressure that occur in the deep veins during muscular contraction, and as a result varices of the saphenous veins occur. Warwick states, furthermore, that the sub-Poupartian valve is absent in a small percentage of subjects. The deep veins may also be inadequately valved below the knee. In one case exhibiting

muscular varicosis he found that "the close valving noted in the posterior tibial venae comites was interrupted over a considerable area, at least two or three inches". He states: "Though the series is not large enough to be conclusive, these experiments suggest that the inadequacy or absence of the valves — more often affecting those in the deep veins, particularly the posterior tibials — is a factor commonly underlying the hereditary tendency to varicosis. This valvular defect is apparently the primary lesion. There is no evidence of weakness in the wall and certainly no sign of dilatation of superficial veins".

Congenital absence of valves above the sapheno-femoral junction has also been pointed out by Eger and Casper (1943) and Eger and Wagner (1949) who, in an attempt to clarify the etiology of varicose veins in the saphenous region, conducted anatomic studies on 38 and 100 cadavers respectively. In the first-mentioned investigation the incidence of congenital absence was found to be 36.8 per cent (bilateral in 7.9 and unilateral in 28.9 per cent, including 10.4 per cent for the right side and 18.5 per cent for the left); in the last-named investigation, the corresponding figure was 42 per cent (bilateral in 6 and unilateral in 36 per cent, including 16 per cent for the right side and 20 per cent for the left). Since the authors had a special problem in mind — the cause of saphenous varices — they did not deal with the distal parts of the femoral vein. They call attention, nevertheless, to the causal significance of the observed absence of valves, pointing out that in the upright position a greater burden imposes on the valves in the femoral and long saphenous veins. Eger and Wagner conclude that "these constitutional defects may explain the hereditary tendency toward the development of varicose veins".

Curtis and Helms (1947), at operation upon a 23-year-old woman who had had an ulcer in the region of the right medial malleolus since the age of 19, found complete absence of venous valves in the external superficial epigastric, pudic branches and upper part of the saphenous veins. They concluded that congenital absence of valves in the larger veins of the femoral and external iliac systems may constitute the underlying cause of varices and leg ulcers. A similar view was propounded by Powell and Lynn (1951) after having conducted an anatomic investigation of the valves of the external iliac, femoral, and upper third of the popliteal veins in 27 cadavers. Their autopsy series included stillborn fetuses as well as adults up to the age of 84 years (*cf.* figure 2). Basmajian (1952) in anatomic dissections of 76 limbs, found total absence of valves in the femoral veins of one case (1.3 per cent).

The first investigator claiming to have demonstrated total congenital absence of valves in the deep veins of the leg seems to have been Luke, who in 1941 described the condition as a new syndrome. His case was a previously healthy woman aged 21, who for one year had suffered from unilateral swelling of the left leg in the afternoons, especially after having been on her feet all day. She gave no familial history of enlargement of the leg. She was married but had had no pregnancies. No varicose or unusually dilated superficial veins were present. Phlebography showed "the absence of, or defective valves in, the deep system of the involved leg, thus allowing complete retrograde flow". The defect was absent in the normal leg. Since, however, Luke states in summing up that "undoubtedly this extremity has been endowed with a set of valves congenitally weaker, and likely numerically fewer, than the normal, and the preceding years

of activity while a waitress produced their complete defectiveness", it seems justified to surmise that his case is an example, not of congenital absence of venous valves but rather of Bauer's pan-phlebois.

In 1951 Luke presented a further three cases, all of them past the age of 20, in which the symptoms had been slight chronic leg enlargement with increased cyanosis and dilated cutaneous venules. Luke wrote: "They had the complaints of aching and a feeling of heaviness and tiredness of the leg when upright for prolonged periods. No edema was present. Retrograde venography gave an identical picture of a valveless common and superficial femoral vein extending, in two cases, into the anterior and posterior tibial branches. The femoral tributaries showed valves but decreased in number. It is my feeling that this is a definite entity: it was described by me in 1941. The leg enlargement is probably due to a congenital absence of valves in deep veins resulting in venous retardation on straining or when upright. It is interesting to note that no cutaneous malnutrition changes were present in the leg in any of the three".

Luke thus considers that these three cases were quite similar to the first he had reported. For this reason, along with the finding of valves in the femoral tributaries — he did not say whether they were competent or not — it is possible that these cases, too, should be assigned to Bauer's pan-phlebois group. However, no pathologic examination of veins was reported in that paper. Another possibility is that the cases could be examples of a variant with a small number of venous valves, for Luke's conclusions indicate that quite a few functioning valves must have been present. "As in saphenous valves", he says, "they (the deep vein valves) can be congenitally weak or absent, but the important point is that such an absence does not lead to the malnutrition changes seen in the valvular incompetence and vein damage of the post-phlebotic leg". — May and Nissl (1959) are of the same opinion when they categorically state, apparently with the support of the case reported by Luke (1941), that they do not believe "*dass lediglich ein kongenitales Fehlen der tiefen Venenklappen zur Entstehung eines ulcus cruris ausreicht*" (that merely the congenital absence of deep venous valves suffices for the development of a leg ulcer.) — To this it may be added that absence of valves, whether congenital or following recanalized thrombosis, would be likely to give rise gradually to similar symptoms of venous insufficiency. Luke's conclusions are quite plausible, however, on the assumption that his cases had functioning valves which escaped observation.

Moore (1951) in a phlebographic investigation of 102 cases with venous insufficiency, found 57 legs without valves in the deep veins. Eighteen of the cases were associated with a history of deep venous thrombosis, and 84 were not. In the remaining 27 cases with no known thrombosis, apparently adequate valves were present. The mean age in Moore's series was 53 and the range 21—66 years. "When valves were absent, the deep vessels were larger than normal. The superficial veins could show good valves, while the deep veins were dilated and showed none". — In view of the high mean age at the onset of symptoms, it seems likely that some at least of the cases were either of pan-phlebois or of recanalization following thrombosis, even though the patients had no histories of thrombosis. Moore himself is evidently inclined to credit the latter of the two possibilities, for he writes in his summary that "two-thirds of the thrombotic legs and all the valveless nonthrombotic legs had a similar

venographic appearance". His view is probably correct, considering the appearance of the shadow produced by a recanalized post-thrombotic vein. According to Bauer (1948) for example, such a vein is phlebographically "irregular, stretches of a very narrow lumen alternating with dilated portions. The contours are rigid and not at all like the soft contours of normal veins".

It should be pointed out in this context that deep venous thrombosis *may* occur without the patient being aware of it. It may happen, for instance, following traumata — not only severe ones such as fractures of the pelvis or of the femoral shaft, but also simple fractures of the fibula and even contusions and abrasions of the extremities (Vance, 1934; Bauer, 1944, and others). Local symptoms such as swelling and pain on palpation make the diagnosis difficult in such cases, and thrombosis, if present, may escape detection. In doubtful cases phlebography may be of great value. Bauer (1944) in phlebographic investigations of 127 cases with deep venous thrombosis, thus found that "thrombosis was twelve times commoner among cases of leg injury than in all the rest of the material".

At ascending phlebography of four males who, around the age of 20, had developed symptoms of venous insufficiency in the lower leg, Norman (1956) was in no case able to identify any valves in the femoral vein proximal to its junction with the short saphenous. He believes that the absence of valves may be congenital and compares his cases to that reported by Luke (1941). However, he also calls attention to the possibility of acquired lack of valves due, say, to undiagnosed mild phlebitis or valvitis in the femoral vein.

Martinet (1959) in a series of 302 unilateral or bilateral retrograde phlebographies (131 static and 171 during straining) in 202 patients with venous insufficiency symptoms, found "*9 Fälle mit Inkontinenz der tiefen Venenstämme bei fehlenden Klappen*" (nine cases with deep venous incompetence due to want of valves). He pointed out that although a congenital valvular deficiency was quite conceivable, the phlebograms in these nine cases had been difficult to evaluate and hence he could not rule out earlier thrombosis, even though there was no history of it.

Lodin, Lindvall and Gentele (1959) reported total congenital absence of valves in all venous regions studied in both the arms and the legs of two sisters (cases 1 and 2 in the present investigation). Each of these patients had developed, around the age of 11, increasing symptoms of venous insufficiency. A few years later, bilateral leg ulcers supervened in both cases. So far as can be judged from my studies of the literature, these are the first reported cases in which total congenital absence of valves in the deep veins of the legs (and arms) has been definitely observed, and this anomaly has thus been differentiated.

CHAPTER 4

PERSONAL INVESTIGATIONS INTO CONGENITAL ABSENCE OF VALVES IN THE DEEP VEINS OF THE LEG

Results and Discussion

The question naturally arose as to the degree in which the relevant valvular anomaly might in itself account for the presence of venous insufficiency in young persons with no record of thrombosis. Furthermore, such cases, in my opinion, were ideally suited for study of the local circulation where pathologic changes had given rise to a lower-leg stasis syndrome. The effect on the general circulation also required elucidation. Patients with congenital absence of venous valves were, as stated, eminently suited for closer scrutiny of these problems, since the general clinical picture was not obscured by previous thrombosis, thrombophlebitis or other local disease processes of the veins.

During 1959 and early 1960, when the series was being collected, I encountered an additional ten cases. All of them sought treatment at outpatient departments of Karolinska Sjukhuset, no fewer than eight at the Department of Dermatology. It is on the total of twelve cases that the present investigation is based. The fact that as many as ten cases were recorded at a single hospital over a period of just over one year indicates that congenital absence of venous valves is not a particularly rare phenomenon.

Table 5. *Age, sex, height, weight, age at menarche, and age at onset of symptoms.*

Case	Age	Sex	Height cm	Weight kg	Age at menarche	Age at onset of symptoms	Age at onset of ulcer
1	32	♀	168	62.2	13	11—12	14
2	21	♀	164	50.0	12	10—11	18
3	41	♀	162	55.5	13	13	27
4	37	♂	176	80.3		15	
5	12	♂	155	40.2		11	
6	9	♀	137	30.4			
7	21	♂	179	68.5		17	18
8	34	♀	171	60.0	13	16	18
9	26	♂	183	69.2		17	17
10	19	♂	184	69.8		15—16	18
11	18	♂	176	63.2		16	17
12	22	♂	178	73.0		16	17

Table 6. *Examinations performed.*

Case	Bacterio- logic cul- tivation of ulcer material	Biopsy of ulcer	Biopsy of vein	Trendelenburg's test						Perthes' test						Oscillography		Skin thermo- metry	Ophthalmologic examination
				Sufficiency		Insufficiency		Unevaluable		Sufficiency		Insufficiency		Unevaluable		at rest	after exercise		
				Right	Left	Right	Left	Right	Left	Right	Left	Right	Left	Right	Left				
1	+		+					+	+							+	+		
2	+	+	+						+	+						+	+		
3			+													+	+		
4			+			+							+			+	+		
5			+				+							+		+	+		
6																+	+		
7															+	+	+		
8	+		+				+					+			+	+	+		
9	+														+	+	+		
10	+		+			+									+	+	+		
11															+	+	+		
12	+	+	+												+	+	+		

Table 7. Roentgen examinations.

Case	Cardio-pulmonary examination	Arteriography of legs		Ascending phlebography of legs		Retrograde phlebography of legs		Pelvic phlebography	Cavo-graphy	Phlebography of arms	
		Right	Left	Right	Left	Right	Left			Right	Left
1	+	+		+	+	+	+	+	+	+	+
2	+	+		+	+	+	+	+		+	
3	+	+	+	+	+	+	+	+		+	+
4	+			+	+	+	+	+		+	+
5	+	+	+	+	+	+	+	+		+	+
6	+			+	+						
7	+			+		+	+	+	+		
8	+	+	+	+	+	+	+	+			+
9	+		+	+	+	+	+	+			
10	+	+	+	+	+	+	+	+		+	
11	+			+	+	+	+	+	+	+	+
12	+	+	+	+	+	+	+	+			+

Before presentation of the individual cases, the composition of the series and the examinations performed will be set forth in tables 5—7. A plus-sign in the tables signifies that the examination in question was carried out.

A. Case Reports

Case 1. Housewife, aged 32. 1074/60.

Heredity. — Parents as well as 8 of 9 siblings (6 sisters and 2 brothers) healthy. One sister (case 2) had recurrent leg ulcers since age 18. Otherwise no known familial history of varices, leg ulcers, other vascular diseases or vascular anomalies.

History. — Had the usual diseases of childhood; otherwise on the whole in good health. No history of thrombosis, thrombophlebitis or varicose veins. No leg injury. Para II.

At age 11—12 (menarche at age 13), onset of increasing edema of the ankles in the afternoons, particularly after standing during the day. The edema subsided nocturnally or when she rested. At age 14 following a slight cut, she had a gradually increasing ulceration at the right medial malleolus. It healed swiftly, however, during hospitalization. Soon after discharge she had a recurrence with several minor ulcers *in loco*. The following year a similar, slow-healing ulcer developed at the left medial malleolus. Over a period of years she was hospitalized several times, the ulcers usually healing quite rapidly with bed rest but returning soon after discharge. Further ulcers developed in due course at the lateral malleoli and on the upper surfaces of both feet. In 1953 resection of the popliteal veins failed to improve the patient's condition. The symptoms were greatly aggravated during pregnancies. She had always suffered from cold hands and feet.

General Findings. — Nothing abnormal. W. R. negative. Conventional urinalysis and blood tests, including serum iron, bleeding time, electrophoretic fractions, etc. normal. Blood pressure 120/80 mm Hg. Mantoux 1 : 1,000 negative. Height 168 cm; weight 62.2 kg.



Fig. 8 a and b. Case 1. The photograph in a was taken in the morning, and that in b six hours later. Note the regular, almost pencil-thick, superficial veins, which in b are practically concealed by the edema that has meanwhile developed.

Local findings (figs. 8 a and b). — On the right medial malleolus was an ulcer measuring 70 by 10—30 mm with a steep, slightly elevated purple rim and a yellowish-red, moist base. The skin around the ulcer and up the lower leg as well as in a wedge-shaped sector extending under the instep was purple, with crust formation in some parts and brown pigmentation with atrophy in others. Around the left medial malleolus was a 15 by 20 mm crusted area surrounded by atrophic purple skin. On each lateral malleolus and each dorsum pedis were ulcer scars. No varicose veins were demonstrable. Moderate edema was present in the feet and over the ankles. Round the ankles and on the upper surfaces of the feet were smooth, almost pencil-thick veins.

Both hands showed highly pronounced erythrocyanosis.

Results of Examinations

Biopsies from the ulcers showed partially necrotic granulation tissue with leukocyte infiltration but no tubercles, giant cells, or other signs of a specific process. No tubercle bacilli were found.

Bacteriologic cultures from the ulcers revealed moderate growth of *Staphylococcus aureus* and enterococci.

Trendelenburg's and *Perthes'* tests were not evaluable.

Pulsations were palpated bilaterally in the femoral, popliteal, posterior tibial, dorsalis pedis, radial and ulnar arteries.

Oscillography (1) at rest and (2) after moderately heavy exercise (5/160) on a staircase: (1) Satisfactory and uniform pulsations in the small of both legs and in both wrists, and (2) normal reaction.

Skin thermometry revealed nothing demonstrably abnormal.

Ophthalmologic examination: N. A. D.; no detectable vascular anomalies in the eyegrounds.

Biopsy of vein. — The small biopsy specimen had a thin intima and well preserved elastic substance in the wall. There was no appreciable fibrosis and no inflammatory cell infiltration.

Roentgenograms of heart and lungs: N. A. D.

Arteriography of right leg: N. A. D.

Phlebography of pelvis and lower extremities. — No signs of valves, either in the deep veins or in the muscle and superficial veins, which were not distended.

Phlebography of arms. — As before, no demonstrable valves either in the deep or the superficial veins, which were not distended.

Case 2. Housewife, aged 21. 1076/60.

Heredity. — See case 1.

History. — Previously healthy on the whole, aside from the usual diseases of childhood. No record of thrombosis, thrombophlebitis or varices. No leg injury. Para I.

At age 10—11 (menarche at age 12) she had increasing edema of the ankles, particularly after prolonged standing. With rest, the edema subsided rapidly and was absent in the mornings. At age 18, onset of ulcerations over both lateral malleoli without known preceding trauma. They healed rapidly with bed rest but soon recurred *in loco*. Similar ulcerations developed in due course round the medial malleoli. During several periods of hospitalization the lesions swiftly healed, but once more recurred shortly after discharge. Edema of the ankles and lower legs was especially pronounced during pregnancy in 1957, when the ulcers were also larger than usual. Patient, like her sister, had always suffered from cold hands and feet.

General findings. — Nothing abnormal. W. R. negative. Conventional urinalysis and blood tests, including serum iron, electrophoretic fractions, bleeding time, etc. normal. Blood pressure 130/75 mm Hg. Mantoux 1 : 1,000 negative. Height 164 cm; weight 50.0 kg.

Local findings. — Just below the right lateral malleolus was a sharply demarcated ulcer 1 by 1.5 cm in size. Around the right medial malleolus and likewise over the malleoli of the left foot, the skin was thin and partially crusted, with brown pigmentation and scars of previous ulcers. No varices were observed. Over the ankles and up the lower legs there was moderate edema. Smooth, almost pencil-thick veins were observed on the upper surfaces of the feet. Both hands showed pronounced erythrocyanosis.

Results of Examinations

Biopsies from the ulcers revealed nonspecific granulation tissue.

In *bacterial cultures* from the ulcers there was growth of *Staphylococcus albus*.

Trendelenburg's and *Perthes'* tests were not evaluable.

Pulsations were palpated bilaterally in the femoral, popliteal, posterior tibial, dorsalis pedis, and the radial and ulnar arteries.

Oscillography (1) at rest and (2) after moderately heavy exercise (5/160) on a staircase: (1) Satisfactory and uniform pulsations in the small of both legs and in both wrists, and (2) normal reaction.

Skin thermometry: Nothing appreciably abnormal.

Ophthalmologic examination: N. A. D.; no demonstrable vascular anomalies in the eyegrounds.

Biopsy of vein. — The small specimen had a thin intima and well preserved elastic substance in the venous wall. It exhibited no appreciable fibrosis and no inflammatory cell infiltration.

Roentgenograms of heart and lungs: N. A. D.

Arteriography of right leg: N. A. D.

Phlebography of pelvis and lower extremities. — No detectable valves in the deep, muscle, or superficial veins, which were not distended except in distal part of the lower legs, where the superficial veins showed slight dilatation.

Phlebography of right arm revealed no valves either in the deep or the superficial veins, which were not distended.

Case 3. Saleslady, aged 41. 1103/60.

Heredity. — Maternal grandfather, one brother and his two children (cases 4—6) suffered from venous insufficiency symptoms similar to those of the patient. Otherwise no known familial history of varicose veins, leg ulcers, other vascular diseases or vascular anomalies.

History. — Aside from the usual diseases of childhood, on the whole in good health. No record of varices, thrombosis, thrombophlebitis or leg injury.

At age 13 (menarche at 13), onset of increasing edema of the ankles and lower legs in the afternoons, particularly after prolonged standing and walking during the day. The edema completely subsided nocturnally or when patient rested. Her condition was more or less unchanged until age 27 when, without any known preceding trauma, stubbornly recurrent ulcerations developed around the malleoli bilaterally; on rare occasions, they also affected the soles of the feet. Numerous types of treatment (elastic bandages, plaster casings, massage, physiotherapy, etc.) produced no lasting improvement. The ulcers usually healed rapidly during hospitalization but recurred shortly after discharge. Patient had always been troubled by cold hands and feet.

General findings. — Nothing abnormal. W. R. negative. Conventional urinalysis and blood tests, including electrophoretic fractions, hemagglutination reaction for rheumatoid arthritis, cold precipitin reaction, cryoglobulins, serum cholesterol, B. M. R., anti-streptolysin and antistaphylococcal titers, etc. were normal. Blood pressure 120/70 mm Hg. Height 162 cm; weight 55.5 kg.

Local findings (figs. 9 a and b). — The skin over the upper surfaces and the malleoli of both feet was thin, reddened, slightly scaling, pigmented brown, and scarred from previous ulcers. Anterior to and below the right lateral malleolus was an ulcer about 2 cm in diameter, with a sharp rim. Both ankles and lower legs showed moderate edema. No varices were discernible. Around the ankles and on the upper surfaces of the feet were uniformly pencil-thick veins.

Both feet and hands exhibited fairly pronounced erythrocyanosis.

Results of Examinations

Trendelenburg's and *Perthes'* tests were not evaluable.

Pulsations were palpated bilaterally in the femoral, popliteal, posterior tibial, dorsalis pedis, and the radial and ulnar arteries.

Oscillography at rest showed satisfactory and uniform pulsations in both calves, both thighs and both wrists, but some discrepancy in the pulsations from the small of the legs — 11 mm for right and 9 mm for left.



Fig. 9 a and b. Case 3. The photograph in a was taken in the morning, and that in b six hours later. Note the edema in b.

Oscillography after moderately heavy exercise (5/160) on a staircase. — Slight insufficiency of the arterial circulation in the small of the left leg, with an inverse recovery phase of under two minutes. For small of the right leg, slightly delayed normal reaction.

Oscillographic evaluation: Slight arterial insufficiency, probably of organic origin, in the left leg. It was unlikely to have had any connection with the venous valvular anomaly other than as a parallel phenomenon.

Skin thermometry revealed nothing demonstrably abnormal.

Ophthalmologic examination: N. A. D.; no detectable vascular anomalies in the eye-grounds.

Biopsy of vein. — The small biopsy specimen showed a thin intima and well preserved elastic substance in the venous wall. There was no appreciable fibrosis and no inflammatory cell infiltration.

Roentgenograms of heart and lungs: N. A. D.

Arteriography of legs: N. A. D.

Phlebography of pelvis and lower extremities. — No valves were detected either in the deep or in the muscle and superficial veins, which were not distended.

Phlebography of arms. — Occasional valves were discerned in the deep veins and solitary ones in the superficial veins of the distal part of the forearms.

Case 4. Accountant, aged 37. 672/59.

Heredity. — See case 3.

History. — Aside from the usual diseases of childhood, patient enjoyed good health on the whole. No record of thrombosis, thrombophlebitis, leg ulcer or leg injury.

At age 15, onset of gradually increasing edema of the ankles and lower legs in the afternoons, particularly after lengthy standing and walking during the day. The edema subsided completely at night or when he rested. Since age 17 he had bandaged his legs with elastic bandages regularly every morning and had worn, throughout the year, high laced boots; these measures had kept the swelling down. He felt that without this bandaging he would have been unable to get along. During the last few years some varicosities had appeared on both lower legs. Patient had always suffered from cold feet.

General findings. — Nothing abnormal. W.R. negative. Conventional urinalysis and blood tests, including electrophoretic fractions, etc. normal. Blood pressure 140/90 mm Hg. Height 176 cm; weight 80.3 kg.

Local findings. — On upper surfaces of the feet, around the ankles and up the lower legs as far as the knees there was fairly pronounced edema. The bandages had produced marked circular impressions, particularly below the knees where they had terminated, on the small of the legs, at the boot tops, and at the base of the toes where the bandaging had started. Both lower legs showed moderate varices. There was no discoloration of the skin, which seemed slightly doughy on palpation.

Results of Examinations

Trendelenburg's and Perthes' tests showed insufficiency.

Pulsations were palpated bilaterally in the femoral, popliteal, posterior tibial, dorsalis pedis, and the radial and ulnar arteries.

Oscillography (1) at rest and (2) after moderately heavy exercise (5/160) on a staircase: (1) Satisfactory, uniform pulsations in the small of both legs and in both wrists, and (2) normal reaction.

Skin thermometry revealed nothing abnormal.

Ophthalmologic examination: N. A. D.; no detectable vascular anomalies in the eye-grounds.

Biopsy of vein. — The small biopsy specimen had a thin intima and well preserved elastic substance in the venous wall. There was no appreciable fibrosis and no inflammatory cell infiltration.

Roentgenograms of heart and lungs: N. A. D.

Phlebography of pelvis and lower extremities. — In the right lower leg somewhat fewer than the normal number of venous valves were observed. The popliteal vein was duplicate, with a functioning valve in one but no valves in the other. Otherwise no valves were detectable in the deep venous trunks except the deep femoral. The superficial veins showed no demonstrable valves; the short saphenous was greatly dilated and emptied into the aulvular one of the two popliteal veins. In the left leg a single valve or so was observed in the distal part of the lower leg; otherwise no valves in the deep venous trunks excluding the deep femoral. No valves were discernible in the superficial veins. The long saphenous vein was somewhat larger in caliber than its counterpart on the right.

Phlebography of arms. — Probably a normal incidence of valves in both the superficial and the deep venous trunks.



Fig. 10. Case 5. Photograph taken at about 3 p.m.

Case 5. Schoolboy, aged 12. 673/59.

Heredity. — See case 3.

History. — Aside from the usual diseases of childhood, enjoyed good health on the whole. No record of thrombosis, thrombophlebitis, varices, leg injury or leg ulcer.

At age 11, onset of gradually increasing swelling of the right ankle and lower leg in the afternoons. After some time the entire leg commenced to swell up as far as the groin, particularly following prolonged standing and walking during the day. The edema completely subsided nocturnally or when patient rested, and was not associated with pain. Elastic bandaging of the whole leg up to the groin reduced the symptoms considerably so that he was able to join in playmates' games without much difficulty. He often played ice hockey. There were no symptoms in the left leg.

General findings. — Nothing abnormal. W. R. negative. Conventional urinalysis and blood tests, including electrophoretic fractions, etc. were normal. Blood pressure 110/80 mm Hg. Height 155 cm; weight 40.2 kg.

Local findings (fig. 10). — The right leg was much thicker than the left. Pressure from the finger on the foot, lower leg and thigh left distinct impressions, indicating edema. No varicosities were detectable. The skin was not discolored. Measurement of

leg circumferences two hours or so after patient had risen from bed in the morning gave the following values:

At level 20 cm above upper border of patella — right 47 cm, left 43 cm.
 At level 10 cm below lower border of patella — right 36 cm, left 31 cm.
 At supramalleolar level — right 25 cm, left 21 cm.

Results of Examinations

Trendelenburg's and *Perthes'* tests were not evaluable.

Pulsations were palpated bilaterally in the femoral, popliteal, posterior tibial, dorsalis pedis, and in the radial and ulnar arteries.

Oscillography (1) at rest and (2) after heavy exercise (10/208) on a staircase:

(1) Satisfactory, uniform pulsations in small of both legs and in both wrists, and (2) normal reaction.

Skin thermometry showed nothing abnormal.

Ophthalmologic examination: N. A. D.; no detectable vascular anomalies in the eye-grounds.

Biopsy of vein. — The small biopsy specimen showed a thin intima and well preserved elastic substance in the venous wall. There was no appreciable fibrosis and no inflammatory cell infiltration.

Roentgenograms of heart and lungs: N. A. D.

Arteriography of legs: N. A. D.

Phlebography of pelvis and lower extremities. — Aside from a solitary valve or two in the principal deep veins of the lower leg, no valves were observed in the deep venous trunks, which were not distended. The short saphenous veins had the normal numbers of valves, but the long saphenous veins could not be evaluated with assurance.

Phlebography of arms. — Probably normal incidence of valves in both the deep and the superficial veins.

Case 6. Girl, aged 9. 1399/59.

Heredity. — See case 3.

History. — Good health, on the whole, aside from the usual diseases of childhood. No record of thrombosis, thrombophlebitis, varices or leg injury.

There had been no symptoms of circulatory disturbance. Patient was examined with respect to heredity.

General findings. — Nothing abnormal. W. R. negative. Conventional urinalysis and blood tests normal. Blood pressure 110/70 mm Hg. Height 137 cm; weight 30.4 kg.

Local findings. — N. A. D.

Results of Examinations

Trendelenburg's and *Perthes'* tests were not evaluable.

Pulsations were palpated bilaterally in the femoral, popliteal, posterior tibial, dorsalis pedis, and in the radial and ulnar arteries.

Oscillography (1) at rest and (2) after heavy exercise (7/160) on a staircase:

(1) Satisfactory, uniform pulsations in the small of both legs and in both wrists, and (2) normal reaction.

Skin thermometry showed nothing abnormal.

Ophthalmologic examination: N. A. D. No detectable vascular anomalies in the eye-grounds.

Roentgenograms of heart and lungs: N. A. D.

Ascending phlebography of legs. — No valves were detected in any part of the deep, nondilated venous trunks in the lower leg and first two-thirds of the thigh; nor were any discerned in the superficial veins except for the right short saphenous, which showed a single valve or so.

Case 7. Typesetter, aged 21. 479/60.

Heredity. — Patient's grandmother, mother, all five maternal uncles, and both maternal aunts had developed varicosities between the ages of about 30 and 40, though none had had leg ulcers. Otherwise no known familial history of vascular diseases or vascular anomalies.

History. — The usual diseases of childhood, but otherwise good health on the whole. No record of thrombosis, thrombophlebitis, or leg injury. Patient was an only child.

Fairly short in stature until approximately age 15, whereafter he grew to his present height within a couple of years. At age 16, began work as hand typesetter, standing most of the day. At age 17, onset of gradually increasing edema of the right ankle and lower leg, particularly marked in the afternoons. The edema subsided completely at night or when he rested. The symptoms gradually increased during the ensuing years, and some varicosities developed on the lower leg, as well as discoloration of the skin over the malleoli. A year or so later there were some minor varicosities together with slight edema of the left ankle in the afternoons. In 1956 (when patient was 18) a slow-healing ulcer developed over the right medial malleolus following a slight abrasion. Phlebography of the right leg in 1958 showed avalvular deep veins without definite distension. In same year patient underwent operation consisting of high ligation, stripping, and local resection of superficial veins in the right leg. The edema thereafter decreased considerably. Patient had always suffered from coldness of the right foot, but had no symptoms in the left leg.

General findings. — Nothing abnormal. W. R. negative. Conventional urinalysis and blood tests, including electrophoretic fractions, etc. normal. Blood pressure 130/80 mm Hg. Height 179 cm; weight 68.5 kg.

Local findings. — Areas of skin the size of the palm, about 10 and 20 cm respectively above the right medial malleolus and on it, were thin, atrophic, brown pigmented, and scarred from previous ulcerations. A similar area about 3 cm in diameter, surrounded by some teleangiectatic zones, was observed at the middle of the posterior surface of the calf. On the upper surface of the foot were several uniformly pencil-thick veins. There was very slight edema of the ankle. The left leg showed a few minor varicosities but otherwise nothing noteworthy.

Results of Examinations

Trendelenburg's and *Perthes'* tests were not evaluable on the right leg. On the left leg Trendelenburg's test showed insufficiency and Perthes' test sufficiency. Pulsations were palpated bilaterally in the femoral, popliteal, posterior tibial, dorsalis pedis and in the radial and ulnar arteries.

Oscillography (1) *at rest* and (2) *after heavy exercise* (5/208) *on a staircase*: (1) Satisfactory, uniform pulsations in the small of both legs and in both wrists, and (2) normal reaction.

Skin thermometry showed nothing abnormal.

Ophthalmologic examination: N. A. D.; no appreciable vascular anomalies in the eyegrounds.

Biopsy of vein. — The small biopsy specimen had a thin intima and well preserved elastic substance in the venous wall. There was no major fibrosis and no inflammatory cell infiltration.

Roentgenograms of heart and lungs: N. A. D.

Phlebography of pelvis and lower extremities. — In the *right* lower leg three valves were discerned in one venous trunk and one valve in another. The femoral vein showed one incomplete valve at the junction with the deep femoral. Otherwise no valves were observed in the deep veins other than the deep femoral. One venous trunk in the lower leg was somewhat distended; the other veins were of normal caliber. The short saphenous vein (1958) was greatly distended and tortuous, and contained no detectable

valves. The long saphenous vein (1958) was narrow and probably normally valved. In the *left* leg, valves were observed in both the femoral and the popliteal veins (lower leg not examined). The short saphenous vein was distended and contained valves only in the distal part of the lower leg.

Case 8. Female telephone operator, aged 34. 1249/60.

Heredity. — Maternal grandfather and his mother had had, and mother and both brothers (cases 9 and 10) had, venous insufficiency symptoms similar to those of the patient; otherwise no known familial history of varicose veins, leg ulcers, other vascular diseases or vascular anomalies.

History. — The usual diseases of childhood. Since 1947 (at age 21) epileptiform seizures in connection with menstruation. During past few years the seizures had been increasingly rare. In other respects patient enjoyed good health on the whole. No record of thrombosis, thrombophlebitis or leg injury.

At age 16 (menarche at 13) varicosities developed fairly rapidly on both legs. A year or so later, onset of increasing edema of the ankles in the afternoons, particularly after prolonged standing and walking during the day. At age 18, with no known preceding trauma, an ulcer developed over the right medial malleolus, then ulcers on the other malleoli. The ulcers periodically healed. At age 24 patient obtained sedentary employment, following which the ulcers healed and she was practically asymptomatic for nine years without treatment. In 1958 (at age 32) an ulcer recurred following a trauma to the left ankle (slight penetrating wound caused by umbrella ferrule). Patient underwent surgery in 1959, with bilateral high ligation, following which she used elastic bandages. The operation had a fairly good initial effect on the symptoms, but the latter subsequently recurred unless she was very careful with the bandaging.

The edema of the ankles and lower legs had increased in the past ten years whenever there had been prolonged standing and walking. It subsided completely at night or when patient rested. She had always suffered from cold hands and feet.

General findings. — Nothing abnormal. W. R. negative. Conventional urinalysis and blood tests, including electrophoretic fractions, etc. normal. Blood pressure 135/80 mm Hg. Height 171 cm; weight 60.0 kg.

Local findings (figs. 11 a and b). — The skin of all the malleoli was thin, atrophic, pigmented brown, and scarred from previous ulcers. Over the left medial malleolus was a sharply demarcated, bean-sized ulcer with a moist surface. There was marked swelling of both ankles and part of the lower legs. No varicosities were detected. Numerous uniformly pencil-thick veins were found on the upper surfaces of both feet.

Results of Examinations

Bacteriologic cultures from the ulcers revealed profuse growth of *Staphylococcus albus*. *Trendelenburg's* and *Perthes'* tests were not evaluable.

Pulsations were palpated bilaterally in the femoral, popliteal, posterior tibial, dorsalis pedis, and in the radial and ulnar arteries.

Oscillography at rest. — The small of the right leg showed normal pulsations and pressure in comparison with both wrists. Pressure and pulsations were somewhat less in the small of the left leg than in the right. Both the right and left calves and thighs showed, however, satisfactory uniform pulsations and satisfactory pressure.

Oscillography after moderately heavy exercise (5/160) on a staircase. — Delayed normal reaction of 1 1/2 minutes for both legs.

Oscillographic evaluation: Borderline values; probably normal conditions.

Skin thermometry showed nothing abnormal.

Ophthalmologic examination: N. A. D.; no demonstrable vascular anomalies in the eyegrounds.



Fig. 11 a and b. Case 8. The photograph in a was taken in the morning, and that in b six hours later. Note the edema which has developed in the interim.

Biopsy of vein. — The small specimen showed a thin intima and well preserved elastic substance in the venous wall. There was no appreciable fibrosis and no inflammatory cell infiltration.

Roentgenograms of heart and lungs. — Findings normal except for left-sided origin of the right subclavian artery.

Arteriography of legs: N. A. D.

Phlebography of pelvis and lower extremities. — In the *right* leg were observed one or two, and in the *left* leg two or three, valves for each deep venous trunk in the middle and distal parts of the lower leg; otherwise no discernible valves in the principal deep veins (aside from the deep femoral veins), which were not distended.

Phlebography of left arm. — One or two incomplete valves were observed in the superficial veins, whereas the deep venous trunks appeared to be normally valved.

Case 9. Male, technician, aged 26. 7078/60.

Heredity. — See case 8.

History. — The usual diseases of childhood but otherwise, on the whole, in good health. No record of thrombosis, thrombophlebitis, or leg injury.

At age 17, onset of increasing swelling around the ankles and up the lower legs, subsequently involving the feet too. After a few months a reddened infiltration about 3 cm in diameter appeared over the left medial malleolus; it developed into a very stubborn ulcer. Edema now extended up to the knees; was more pronounced after prolonged standing and walking during the day, but subsided completely at night or when patient rested. At age 18, very painful ulcers periodically on the dorsal surfaces of the left first to third toes. Patient was hospitalized in 1952 with diagnosis of suspected Buerger's disease. — *Oscillometry* and *skin thermometry* showed nothing abnormal.

Oscillography at rest: Normal findings.

Oscillography after moderately heavy exercise (5/160) on a staircase, as well as *elevation test*: Pathologic results for left leg. Investigation revealed no definite evidence of Buerger's disease. The peripheral arteritis was considered to be probably of infectious etiology. In the hope of improving the skin circulation, *left lumbar sympathectomy* was resorted to in 1953, but produced no amelioration. Slow-healing recurrent ulcers were found over the left medial malleolus and the toes a week or so after patient's discharge; subsequently there were repeated recurrences. Patient had his legs bandaged during 1954—1955, and the ulcers then healed. In 1955 a chronic ulcer developed over the right lateral malleolus. During the past year there had been dry, desquamative lesions on the distal part of the right dorsum pedis. There was no pain in the legs even after long walks. The leg edema remained unchanged. Since age 19—20, increasing varicosities of both legs. Patient had always suffered from cold hands and feet.

General findings. — Nothing abnormal. W. R. negative. Conventional urinalysis and blood tests, including serum cholesterol, absorption reaction for rheumatoid arthritis, blood calcium, antistreptolysin and antistaphylococcal titers, etc. were normal. Blood pressure 125/80 mm Hg. Mantoux 1 : 100 positive. Height 183 cm; weight 69.2 kg.

Local findings (fig. 12). — An area of purple skin, the size of the palm, over the left medial malleolus was pigmented brown in parts and scarred in parts. A sharp-rimmed ulcer with a moist surface and 2.5 by 3 cm in size was present at its center. Across the first to fourth toes and extending like a wedge over the ankle was an area of thin purple skin containing scars from previous ulcers. Similar lesions extended across the right lateral malleolus, and at the center was a sharply demarcated ulcer 2 by 4 cm in size, with a gelatinous surface. The distal part of the right dorsum pedis showed increased pigmentation and dry scales.

There was fairly marked edema of both feet and lower legs. The latter showed pronounced varicosities, and the upper surfaces of the feet smooth, uniformly pencil-thick veins.

Results of Examinations

Bacteriologic cultures from the ulcers revealed profuse growth of *Proteus vulgaris*.

Trendelenburg's and *Perthes'* tests showed insufficiency.

Pulsations were palpated bilaterally in the femoral, popliteal, posterior tibial, dorsalis pedis and in the radial and ulnar arteries.

Oscillography (1) *at rest* and (2) *after moderately heavy exercise (5/160) on a staircase*: (1) Satisfactory, uniform pulsations in the small of both legs and in both wrists, and (2) normal reaction.

Skin thermometry showed nothing worthy of note.

Ophthalmologic examination: N. A. D.; no demonstrable vascular anomalies in the eyegrounds.

Roentgenograms of heart and lungs: N. A. D.

Arteriography of left leg: N. A. D.

Phlebography of pelvis and lower extremities. — In the distal two-thirds of the lower legs, two or three valves per deep venous trunk were observed on the right side



Fig. 12. Case 9. Note the lesions resembling those in Buerger's disease, at the base of the toes.

and three or four on the *left*. Otherwise, aside from the deep femorals, no valves could be discerned in the principal deep veins, which were not distended. The right long saphenous vein showed one incompetent valve at the junction with the femoral vein; otherwise there were no detectable valves in the greatly distended superficial veins.

Case 10. Factory worker, male, aged 19. 1275/60.

Heredity. — See case 8.

History. — The usual diseases of childhood; otherwise healthy on the whole. No record of thrombosis, thrombophlebitis or leg injury.

At age 15—16 patient was distressed by gradually increasing sensation of fatigue in the legs, as well as edema of the ankles and lower legs in the afternoons, particularly after prolonged standing or walking during the day. The edema disappeared nocturnally or when he rested. At age 18, a few small varices developed on anterior surfaces of the lower legs; injection treatment one year later had good results. Also at age 18, onset of ulcerations on both medial malleoli without known preceding trauma; the ulcers grew somewhat in size but at no time healed. Patient had always complained of cold hands and feet. He had never bandaged his legs.

General findings. — Nothing abnormal. W. R. negative. Conventional urinalysis and blood tests, including electrophoretic fractions, antistreptolysin and antistaphylolysin titers, etc. normal. Blood pressure 125/80 mm Hg. Height 184 cm; weight 69.8 kg.

Local findings. — At the left medial malleolus was a sharp-rimmed ulcer, 4 by 4 cm in size and in part crusted, surrounded by a fairly large area of thin, brown-pigmented skin. Similar lesions were observed over the right medial malleolus, with a bean-sized ulcer and scars from previous ulcerations. There was moderate swelling of the ankles and parts of the lower legs. At the middle of the medial surface of the left lower leg were a few very slight varicosities. Uniformly pencil-thick veins were noted on the upper surfaces of both feet.

Results of Examinations

Bacteriologic cultures from the ulcers showed profuse growth of *Staphylococcus albus*. *Trendelenburg's* and *Perthes'* tests were not definitely evaluable.

Pulsations were palpated bilaterally in the femoral, popliteal, posterior tibial, dorsalis pedis and in the radial and ulnar arteries.

Oscillography (1) at rest and (2) after heavy exercise (10/208) on a staircase: (1) Satisfactory, uniform pulsations in the small of both legs and in both wrists, and (2) normal reaction.

Skin thermometry showed nothing noteworthy.

Ophthalmologic examination: N. A. D.; no detectable vascular anomalies in the eye-grounds.

Biopsy of vein. — Biopsy specimen was small and in the lumen was found an organized, recanalized thrombosis residue. Elastic substance of the wall was but little changed. Venous wall possibly was slightly sclerosed. No appreciable inflammatory cell reaction.

Roentgenograms of heart and lungs: N. A. D.

Arteriography of legs: N. A. D.

Phlebography of pelvis and lower extremities. — In the right lower leg were observed one or two valves per venous trunk; in the left lower leg a solitary valve or so. There was a single valve or two peripherally in the right deep femoral vein; the left deep femoral had a normal number of valves. Otherwise, no discernible valves either in the deep venous trunks, which were not distended, or in the superficial veins.

Phlebography of right arm. — A solitary incomplete valve or so was detected in the superficial veins, whereas the deep veins appeared to have a normal number of valves.

Case 11. Male student, aged 18. 962/59.

Heredity. — No known familial history of varicosities, leg ulcers, other vascular diseases or vascular anomalies.

History. — Previously healthy on the whole, aside from the usual diseases of childhood. No record of thrombosis, thrombophlebitis or leg injury.

In 1957 at age 16, onset of increasing swelling of the left ankle in the afternoons, and in the following year similar symptoms on the right side. There was a short period of slight bilateral varicosities, which subsided completely after patient had worn elastic stockings for a time. The edema of the legs had gradually increased during the last few years. It subsided rapidly with rest, and by the mornings had quite disappeared. Since 1958 there had been slow-healing, recurrent ulcers of the malleoli and on the upper surfaces of the feet at the shoe line. They healed swiftly with bed rest, but recurred when patient was ambulatory. The symptoms gradually subsided if he wore elastic stockings regularly, and periodically he was almost free of symptoms. He had always suffered from cold hands and feet.

General findings. — Nothing abnormal. W. R. negative. Conventional urinalysis and blood tests, including electrophoretic fractions, liver function, L. E. cells, antistreptolysin

and antistaphylolysin titers, electrocardiograms, etc. normal. Blood pressure 130/65 mm Hg. Height 176 cm; weight 63.2 kg.

Local findings. — On the right dorsum pedis were a few discrete ulcers two centimeters or so in diameter. Similar ulcers were observed on the anterior surface of the right lower leg approximately 10 cm above the ankle, as well as over the left medial malleolus. The skin of the ankles and malleoli was thin, pigmented brown, and scarred from previous ulcerations. On the upper surfaces of the feet and across the ankles were numerous uniformly pencil-thick veins. There was fairly pronounced edema over the ankles and some distance up the lower legs. No varicosities were detected.

Results of Examinations

Trendelenburg's and Perthes' tests were not evaluable.

Pulsations were palpated bilaterally in the femoral, popliteal, posterior tibial, dorsalis pedis and in the radial and ulnar arteries.

Oscillography (1) at rest and (2) after heavy exercise (10/208) on a staircase: (1) Satisfactory, uniform pulsations in the small of both legs and in both wrists, and (2) normal reaction.

Skin thermometry showed nothing noteworthy.

Ophthalmologic examination: N. A. D.; no detectable vascular anomalies in the eye-grounds.

Roentgenograms of heart and lungs: N. A. D.

Phlebography of pelvis and lower extremities. — Aside from two peripherally located valves in the right and one in the left deep femoral vein, no valves were observed either in the deep or the muscle or superficial veins, which were not distended. (The possibility of one or two isolated valves in the minor superficial veins was not entirely precluded.)

Phlebography of arms. — A single valve or two was observed in the deep veins of the left forearm; otherwise no valves were discernible in either the deep or superficial veins.

Case 12. Mechanic, aged 22. 536/60.

Heredity. — Father and paternal grandmother had varicose veins. Father suffered from repeated thrombophlebitis and attacks of pain in the legs, which had been regarded as thrombosis. Phlebography in 1960 showed, however, a normal incidence of valves in both legs. Otherwise, no familial record of varices, leg ulcers, vascular diseases or vascular anomalies.

History. — Patient had had the usual diseases of childhood. Aside from surgery for left inguinal hernia in 1944, he had enjoyed good health on the whole. No record of thrombosis, thrombophlebitis, or leg injury.

At age 16 an exhausting spell of exercise (running barefoot in a gymnasium for one hour) had been followed, the same evening, by pronounced swelling of the feet and lower legs which made it difficult to remove his shoes. After a day or so in bed the swelling had disappeared. Subsequently there was periodical discomfort from swollen ankles, particularly after prolonged standing or walking during the day. The swelling disappeared at night or when he rested. At age 17 without any known preceding trauma, an ulcer developed on the left medial malleolus and at no time thereafter showed complete healing. Somewhat later another ulcer appeared on the lateral malleolus. Patient was admitted several times to his local hospital, but no reliable diagnosis was established. In 1956, *biopsy* from the ulcers showed non-specific granulation tissue. *Bilateral arteriography* of the legs in 1957 showed a normal vascular anatomy. Phlebography the same year revealed no circulatory obstruction in the deep veins of both legs, which were of normal caliber and appearance.

A few minor varicosities on the posterior surface of the left calf led, in 1959, to

low ligation in that leg, chiefly in the hope of promoting healing of the ulcers. The operation, however, produced no improvement. Patient had always suffered from cold hands and feet.

General findings. — Nothing abnormal. W. R. negative. Conventional urinalysis and blood tests, including electrophoretic fractions, cryoglobulins, antistreptolysin and antistaphylococcal titers normal. Blood pressure 120/80 mm Hg. Mantoux 1 : 2,000 negative. Height 178 cm; weight 73.0 kg.

Local findings. — In an area double the size of the palm, over and behind the left medial malleolus, the skin was thin, brown-pigmented, desquamative and partially crusted. In the center of this area was a fairly deep, sharply defined ulcer 3 by 4 cm in size, with a moist surface. At the lateral malleolus were similar lesions and two ulcers about 1.5 cm in diameter.

No varicosities were detectable aside from a few very slight ones. Moderate edema was present over the ankles. Uniformly pencil-thick veins were observed on the upper surfaces of both feet and on the lower legs, particularly the right. The skin of both feet showed fairly pronounced cyanotic discoloration. Otherwise the right leg seemed normal.

Results of Examinations

Bacteriologic cultures from the ulcers revealed growth of *Staphylococcus albus*.

Trendelenburg's and *Pertbes'* tests were not evaluable.

Pulsations were palpated bilaterally in the femoral, popliteal, posterior tibial, dorsalis pedis, and in the radial and ulnar arteries.

Oscillography at rest showed satisfactory and uniform pulsations in both legs and both arms.

Oscillography after moderately heavy exercise (5/160) on a staircase. — Delayed normal reaction of 1 1/2 minutes for small of the right leg, and inverse recovery phase of 2 minutes for small of the left leg.

Oscillographic evaluation. — The right and left legs differ in that the latter shows an inverse recovery phase of 2 minutes. This may well point to slight arterial insufficiency, and arteriography is recommended. A similar finding might be recorded if the vessels were of finer caliber in the left leg, and it does not necessarily signify a local obstruction.

Skin thermometry. — Temperature fall of 10° C for the left leg and 13° C for the right. Comparison showed that the temperature for the right first to third toes was not more than approximately 20.2° C but for the corresponding left toes was 23.4° C. This discrepancy appeared to exceed the normal variation. The left fingers were somewhat colder than the right, the difference being around 2° C. Otherwise skin temperature measurement on the arms showed nothing noteworthy.

Ophthalmologic examination: N. A. D.; no detectable vascular anomalies in the eye-grounds.

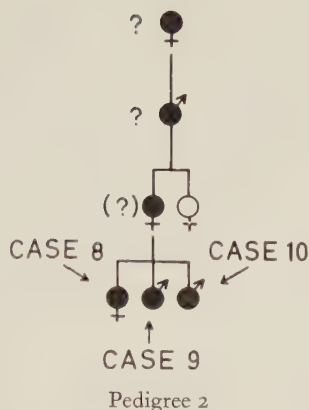
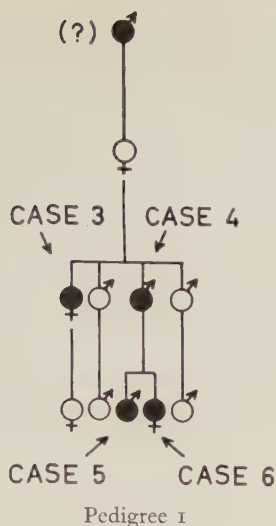
Biopsy of vein. — In the lumen of the small biopsy specimen was an organized, recanalized thrombosis residue. The elastic substance of the wall was little changed. Possibly the venous wall was slightly sclerosed but there was no appreciable inflammatory cell reaction.

Roentgenograms of heart and lungs: N. A. D.

Arteriography of legs: N. A. D.

Phlebography of pelvis and lower extremities. — One incompetent valve was discerned in a branch of the right deep femoral vein distally. In the distal parts of both lower legs an incomplete valve or two was observed; otherwise, no detectable valves either in the deep venous trunks or the muscle veins, which were not distended. Both the femoral and the popliteal veins in each leg were duplicate.

Phlebography of left arm showed normal incidence of valves.



B. Heredity

Pedigrees 1 and 2 show that congenital absence of venous valves or symptoms commonly associated therewith can be traced back for four generations in some families. Bearing in mind the comparative rarity of the phenomenon, this finding may be said to point to a hereditary predisposition.

As regards pedigree 1, the children of the male ancestor born in 1853, said that he suffered from swollen legs since early youth (probably since the age of 13—14). The swelling was similar in type to that in cases 3—6; i. e., it subsided nocturnally and with rest, but returned during the day. He had not suffered from varicose veins or any known thrombosis. It was his custom always to bandage his legs with some stout material and to use high laced boots (*cf.* case 4). If he neglected these measures the swelling of the legs increased and ulcerations developed at the malleoli. An ulceration of this kind first appeared when he was around the age of 20. Although there is no conclusive evidence that this man suffered from congenital absence of venous valves, it seems highly plausible to assume that he did so, for the clinical picture closely resembles that of other members of his family who manifested this anomaly. It may be mentioned that in cases 3 and 4, the parents and a maternal aunt, as well as the daughter of case 3, showed phlebographically a normal incidence of valves in the deep veins of the legs.

In the case of pedigree 2 as well, venous insufficiency was traced back for four generations. The female ancestor born in 1841 was known to have had bilateral leg ulcers before the age of 20, without any record of thrombosis or thrombophlebitis. Probably she had not suffered from varicose veins. A unipara, she had borne a son at the age of 28. Her husband enjoyed good health. The son, who suffered neither from thrombosis nor thrombophlebitis or varicosities, developed symptoms of venous insufficiency in his early youth, and acquired bilateral leg ulcers even before the age of 17. His wife was in good health. — In neither of these two instances can it be positively stated that the venous

insufficiency had been due to congenital absence of valves. However, there are strong grounds for the assumption that such was the case, considering the clinical similarities between these subjects and the avalvular patients.

In generation 3 the mother, aged 59, had since the age of 16—17 (menarche had occurred at age 16) suffered from increasing swelling of the ankles and lower legs, particularly after prolonged standing or walking during the day. The edema subsided completely at night and on resting. At age 20 she had developed an ulceration on the right medial malleolus following a superficial abrasion caused by a shoe heel. Twelve years later a similar ulceration had appeared spontaneously at the left medial malleolus, followed, soon afterwards, by additional ulcerations on the lateral malleoli. She had been hospitalized several times with various diagnoses. The ulcerations healed rapidly during bed rest but invariably recurred shortly after discharge. The superficial veins of the ankles and upper surfaces of the feet had always been almost pencil-thick and regular. She had not previously suffered from varicose veins, but after age 50 developed progressive varicosities of the lower legs. She had a life-long record of cold feet.

At the age of 36 she acquired erysipelas of the right leg, developing from the ulceration at the medial malleolus. While confined to bed for this condition, thrombosis of the leg supervened. Phlebography in 1960 disclosed two incompetent valves on either side in the proximal portions of the femoral veins. The upper one of the two on the left side was incompletely developed. Otherwise no venous valves were observed in the deep venous trunks of the leg, which were not dilated. Evaluation of the phlebograms was difficult, however, due to the numerous superficial varicosities. In the distal part of the right popliteal vein were observed lesions suggestive of recanalization following thrombosis. The roentgenologic findings were not inconsistent with the possibility of congenital absence of valves in the deep veins distal to the upper sectors of the femoral veins. Phlebography of the left arm revealed a normal incidence of valves, and roentgenograms of the heart and lungs showed nothing abnormal.

The history in this case closely accorded with that in the other patients with congenital absence of valves. It is probable, therefore, that she suffered from a like congenital anomaly. However, in view of the thrombosis and the profuse varicosities which had developed during the past few years, the possibility that the venous insufficiency had some other etiology is not altogether precluded.

It should be pointed out that her three children had different fathers, all of whom were, as was her sister, free from leg symptoms. It may thus be concluded that the chance of congenital absence of venous valves is greater than average for the offspring of a person with that anomaly.

The fact that congenital absence of valves may be due to an hereditary predisposition is perhaps not surprising; for it has long been recognized that other venous lesions such as varicosities, show a marked familial tendency. Thus Magnus (1921) reported that between 50 and 75 per cent of his patients with varicose veins gave a history of an hereditary factor, while of those studied by De Takats and Quint (1930) 65 per cent reported such a factor. Foote (1950) calls attention to the fact that "not only is the hereditary tendency present, but it is not uncommon to see the same pattern of varices transferred from

parent to child. Again, not only may the pattern be transferred, but the type of vein itself may run in the families". On analyzing the hereditary factor in 525 patients, Anning (1950) noted a familial history of ulcers or varicose veins in 256 (51.6 per cent). Dodd and Cockett (1956) observed, in 100 consecutive cases of varicose veins, a corresponding figure of 79 per cent, whereas Sigg (1958) reports an incidence as high as 85 per cent.

If, therefore, a heredity may exist in which weakness of the veins leads to varicosities, it is not surprising that variations in the number of venous valves, including total absence thereof, may be attributable to a hereditary factor.

It was pointed out in the foregoing that the number of venous valves may vary substantially in different individuals and still be within the "normal range". A large number of variants, ranging from profusion to complete absence of valves, probably exists. An example of *one* variant is the following case:

Housewife, aged 23.

Heredity. — Maternal grandmother had had leg ulcers late in life. Mother, who was uniparous, had varicosities in connection with pregnancy. Otherwise there was no familial history of varices, leg ulcers, vascular diseases or vascular anomalies.

History. — Suffered from the usual diseases of childhood, but otherwise enjoyed good health aside from bronchial asthma stemming from sensitivity to dog. There was no record of thrombosis, thrombophlebitis or leg ulcer.

As far back as she could remember, she had suffered from acrocyanosis of the hands and feet in cold weather. The veins on her legs had always been large, regular and conspicuous. In connection with menarche at age 13 several small teleangiectatic areas had appeared on the feet. There was a tendency to nocturnal cramps in the calves, particularly during menstruation. Since age 16—17, patient had experienced increasing sensation of muscle tension in the calves and feet, especially after prolonged standing — as well as swelling of the ankles and lower legs, particularly the left. During the past few years there had been increasing, discolored teleangiectatic areas on the upper surfaces of both feet and round both ankles, together with numerous regular, distended, pencil-thick veins. A few small varicosities had appeared on the posterior surface of the left calf in the last 12 months or so. Patient underwent *Cockett's operation* (division of communicating veins) in October 1959, after which her condition was improved insofar as both the distended venous trunks and the teleangiectatic areas decreased substantially. The discomfort was then only slight, though she continued to wear elastic stockings.

General findings. — Nothing abnormal. W. R. negative. Conventional urinalysis and blood tests normal. Blood pressure 130/70 mm Hg. Mantoux: 1 : 100 positive. Height 180 cm; weight 60.0 kg.

Local findings. — Moderate edema of the feet and over the ankles. Around the ankles and on the upper surfaces of the feet were veins of uniform pencil thickness. No varicosities were detectable except for a few slight ones on the posterior surface of the left calf. There were no ulcers or ulcer scars. The skin of both feet showed moderate cyanotic discoloration.

Results of Examinations

Trendelenburg's and *Perthes'* tests were not evaluable.

Pulsations were palpated bilaterally in the femoral, popliteal, posterior tibial, dorsalis pedis and in the radial and ulnar arteries.

Oscillography at rest showed satisfactory, uniform pulsations in the calves, thighs, upper arms and forearms.

Skin thermometry revealed nothing abnormal.

Ophthalmologic examination: N. A. D.; no detectable vascular anomalies in the eye-grounds.

Roentgenograms of heart and lungs: N. A. D.

Phlebography of pelvis and lower extremities (ascending and retrograde). — Both lower legs showed a normal incidence of valves; the muscle veins and deep femoral veins were also valved. No valves were detected, however, either in the popliteal veins or more proximally in the deep venous trunks (though the distal part of the right femoral vein could not be satisfactorily evaluated). Both short saphenous veins were dilated and contained no demonstrable valves.

C. Sex Distribution

It is evident from the foregoing that the anomaly appears to be equally common in males and females. The present series is too small, however, to permit any definite conclusions in this respect.

D. Age at Onset

It is difficult to determine the precise time at which clinical signs of venous insufficiency first appear. Edema of the legs, increasing during the day and heightened by prolonged standing and walking, is said by most authors to be the earliest and also the most frequent symptom of chronic venous insufficiency. Piulachs (1956), for instance, writes that "unlike nearly all the rest of the sequels (except pain), it (the venous insufficiency) does not develop after a long period, but appears at an *early* date". All of the patients in the present series (except case 8) reported increasing swelling of the ankles and/or lower legs as the initial symptom. Most of them said the onset had occurred around the age of puberty. Hence it should be justifiable to regard that as the usual age of onset. It would seem plausible, moreover, to infer at least a partial connection between the initial symptoms and the coincident growth in height. Due to the absence of valves the veins in the lower part of the body are like open tubes whose length roughly coincides with the distance between heart level and the dorsum pedis, the result being a high hydrostatic pressure on the tissues of the lower leg as long as the subject is erect. With growth in height, this pressure increases. Persons with congenital absence of venous valves are in an even worse situation than are those with post-thrombotic conditions, since all the deep veins of the lower legs are (more or less) avalvular. The orthostatic pressure acting upon the whole of the lower leg is probably a factor in the early onset of symptoms. In this connection numerous similarities may be found with the conditions associated with varicose veins, since according to Eger and Wagner (1949) the following factors are of significance as respects the fact that varicose veins rarely develop before adolescence:

1. Elasticity of veins and skin is greatest then.
2. Diameter of veins is relatively smaller.
3. Veins are still growing in length, minimizing tortuosity.

4. Hydrostatic pressure is less, since full length has not been attained.
5. Muscular movements are more active and more constant.
6. Prolonged periods of standing, occupational or otherwise, are exceptional”.

It should be made clear that none of the patients in the present series was adipose; on the contrary, the subjects tended to be below normal weight. Case 4 had put on some weight during the past few years, one of the reasons probably being that his symptoms had necessitated a more sedentary way of life.

E. Symptoms and Course

By and large, the symptoms and course conformed with those found in venous insufficiency of other etiology. The leg ulcers, for instance, had the classical malleolar localization. One discrepancy, however, was the early age at onset in most of the cases. Moreover, varicosities usually did not occur initially but developed after several years — and then in only a few cases. However, slight dilatation of the superficial veins, particularly on the upper surfaces of the feet, was found in all cases. The deep veins were visualized phlebographically as straight and regular, and perhaps somewhat narrower than normal. Furthermore, both the development and the subsidence of leg edema occurred more rapidly than is customary in venous insufficiency of other etiology. — The clinical symptomatology alone, however, is not sufficiently characteristic to establish the diagnosis; the latter must be verified by phlebography.

F. Leg Injury

The patients' histories were meticulously recorded with special reference to leg injuries. None of the patients had knowledge of any such injury. Although thrombosis may follow even minor traumata, these patients are most unlikely to have had thrombosis with recanalization in all deep veins of the leg. Nor did the phlebographic findings resemble those observed in post-thrombotic conditions. Hence the supposition that the absence of valves might have been due to earlier thrombosis can be practically ruled out.

G. Other Vascular Anomalies

Only in one patient (case 8) was another vascular anomaly demonstrable; namely, left-sided origin of the right subclavian artery. An anomalous subclavian artery of this character is not, however, altogether unusual: Felson *et al.* (1950) in their studies of the literature found that various authors had reported incidences of 0.4 to 2.0 per cent in autopsy series. The anomaly in the present case might therefore be regarded as an incidental finding, unrelated to the absence of valves in the deep veins of the leg.

In one patient (case 4) the right popliteal vein was duplicate; in another (case 12), both the femoral and popliteal veins were bilaterally duplicate. Duplication of these veins is, however, so common — “reduplication of venous

channels in the popliteal fossa and Hunter's canal is thus the rule rather than the exception", according to Dodd and Cockett (1956) — that the anomaly in these two cases may well be regarded as incidental and without relation to the absence of venous valves.

The arms were phlebographed in nine patients. Venous valves were totally absent in two cases and partially absent in two; in two others they were partially absent only in the superficial venous system of the arms. In three the number of valves was normal.

As regards the normal incidence of venous valves in the arms, Poirier (1896) points out that valves are more numerous in the deep than in the superficial veins, Houzé (1854) having recorded an incidence of one valve per 3 or 4 cm in the deep veins as compared with one per 4 or 5 cm in the superficial veins. Poirier goes on to give the following figures: five to fifteen valves for the brachial vein; eight to twelve for the radial; up to fifteen for the ulnar; six or seven for the forearm portion and four to eight for the upper-arm portion of the basilic; four to seven for the forearm portion and eight for the upper-arm portion of the cephalic, three of the latter eight being in the deltoideus part and one valve at the junction with the axillary vein. The medial basilic vein may be either valved or avalvular, whereas the median cephalic is invariably valveless.

In no case were *vascular anomalies* observed in the eyegrounds. The examinations were performed by an ophthalmologist and in all cases resulted in normal findings.

H. Arterial Circulation of the Extremities

Investigation of the arterial circulation in the extremities is advisable not only to rule out arterial abnormalities as etiologic factors but also in view of the possible existence of arterial anomalies as a parallel phenomenon.

In none of the present cases was the arterial system of the extremities found to show any major abnormalities. Palpation of the major arteries (femoral, popliteal, posterior tibial, dorsalis pedis, radial and ulnar) invariably disclosed satisfactory and uniform pulsations.

For objective verification of the palpatory findings and for the purpose of securing further information, the patients underwent, at Ejrup's laboratory, *oscillography at rest and after exercise* as well as *skin thermometry*. For the technique and fields of use in these examinations, see Ejrup's works (1948, 1949, 1960).

It must suffice here to mention briefly that the oscillographic examinations were performed before and immediately after running on a staircase. The skin temperature, which reflects the blood circulation rate in the skin, was measured after the patients had reclined with extremities exposed for 30 minutes. Two consecutive thermometric determinations were made on the toe tips and finger tips as well as on the arms and legs with the aid of a template. To insure determination in exactly the same area both times, small crosses were drawn on the skin and the electrode applied at the center of each.

In three patients (cases 3, 8 and 12) a slightly delayed recovery phase was recorded after exercise oscillography. Although this finding probably had no

etiologic connection with the symptoms of venous insufficiency, a certain discrepancy between arterial supply and muscular requirement may naturally have existed. Such a discrepancy might suggest that these three patients had poorer vascular systems in general. It may be pointed out, however, that none of them exhibited symptoms of other diseases which, according to Ejrup (1952 a, 1952 b), may be associated with a delayed recovery phase, as for instance peripheral organic arterial stenosis, coarctation of the aorta, decompensated heart diseases, grave anemia, disturbance of the calcium metabolism, and diabetes. Furthermore, a normal arterial anatomy of the legs was verifiable arteriographically both in these three patients and in the others who were examined. Arteriography, unilateral or bilateral, was performed in eight of the cases.

In summing up his reports Ejrup observes that in all cases except the above-mentioned three borderline cases, the examinations "have yielded no evidence of organic obliterative lesions of the arteries of the arms or legs". Even the three borderline cases were, arteriographically, normal. It seems most unlikely, therefore, that the changes in the arterial circulation of the legs had any causal relationship to the patients' symptoms. Nor does there appear to be any support for the assumption that arterial anomalies might be more common, as parallel phenomena, in persons with congenital absence of valves in the deep veins of the leg than in others.

I. Venous Circulation of the Legs

In the opinion of most authors today, venous insufficiency is one of the sequels of venous stasis. Bauer (1947), for instance, writes: "It seems to have been established that all leg ulcers, excluding the purely specific ones, as well as the varicosities and indurations that accompany them are caused by venous stasis, i. e. an impeded venous return from the area in which the ulcer is situated. The fundamental cause is, as a rule, insufficiency of some important sector of the venous return from the lower leg". — The cases reported in this paper might well be considered to typify such venous insufficiency, since they had few or no venous valves in the legs. It should, therefore, be justifiable to regard them as probable examples of venous insufficiency as a sequel of impaired venous return.

Of importance in this context is the fact that one of the patients (case 7) was, in all probability, devoid of valves only in one leg, which moreover exhibited symptoms of venous insufficiency. The contralateral symptom-free leg showed a normal number of valves in the deep veins of the thigh. Unfortunately this patient refused to undergo phlebographic examination of the asymptomatic lower leg. It seems probable, however, that the deep veins therein were normally valved, for not only were symptoms of venous insufficiency absent, but Perthes' test showed sufficiency.

According to Brecher (1956), "the essential function of the venous valves is to break up the high hydrostatic fluid column into small segments so that the venous reservoir is not overfilled by the full pressure of the uninterrupted column. When the lower venous segment is locally compressed by a muscle contraction, pressure in this segment briefly rises above that in the higher seg-

ment and flow is established. When valves are incompetent a high fluid column over-distends the veins, and due to a high-pressure (filtration) gradient across the venous and capillary walls, orthostatic edema may ensue". — Patients with total absence of valves must, as mentioned above, be regarded as clear-cut examples of such conditions. In the erect position the muscle pump of the leg cannot function satisfactorily since with every muscle contraction the blood is also forced in a retrograde direction, the result being a temporary increase of pressure. Piulachs (1956) likens this retrograde transmission of the hypertensive waves to blows of the venous hydraulic ram that gives rise to lesions of the smaller vessels. The venous blood flow is slower than that in the normal leg, and increasing edema develops. A series of lesions is produced, which are attributed to anoxia; the traumatic effects on the tissues eventually result in ulcerations. Problems in this field will be further studied in the clinico-physiologic investigations that are now in progress and whose aim is, if possible, to enhance our understanding of venous insufficiency in the lower leg.

J. Histopathologic Examination of Venous Biopsy Specimens

Bauer has studied a large number of venous biopsies. His concept of pan-phlebotis has already been described in this paper. In Bauer's view the morbid anatomy of pan-phlebotis is similar in all patients, irrespective of the venous zone from which the specimens have been taken. The lesions in Bauer's cases included marked fibrosis and splitting of the elastic elements. He also points out that the phlebo-sclerotic process is situated largely in the intima.

In the present investigation venous biopsies were examined histopathologically in nine of the cases. Small specimens of vein were excised from the dorsum pedis. Seven of these patients exhibited a histologic picture fully in accord with that found in normals of corresponding age. The intima was thin, the elastic substance in the venous wall well preserved, and appreciable fibrosis lacking (figures 13 and 14). In the other two patients (cases 10 and 12), the small veins removed from the dorsum pedis showed organized, recanalized thrombosis residue. Microscopic examination otherwise revealed, however, only very slight changes in the walls of these vessels, the elastic substance of which was well preserved.

In short, therefore, lesions of the type considered by Bauer to be indicative of pan-phlebotis were not encountered in any of the present cases. The logical conclusion, therefore, is that these cases cannot be classed along with those described by Bauer under the designation of pan-phlebotis.

K. Therapy

No causal therapy proper exists for post-thrombotic conditions, nor for congenital absence of venous valves. Several extensive investigations have been reported in regard to conservative treatment of venous insufficiency (Anning, 1954; Foote, 1954; Dodd and Cockett, 1956; Rastgeldi, 1957; Gilje, 1958; and others). It is evident from case 4 that such treatment, if persevered with, may

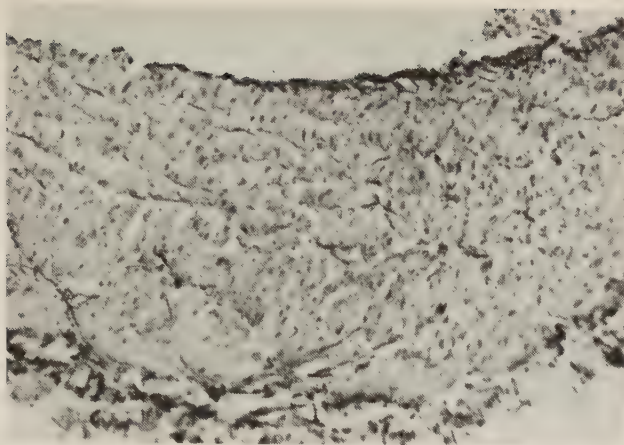


Fig. 13. Case 2. Photomicrograph of venous biopsy. Elastin. $\times 100$.

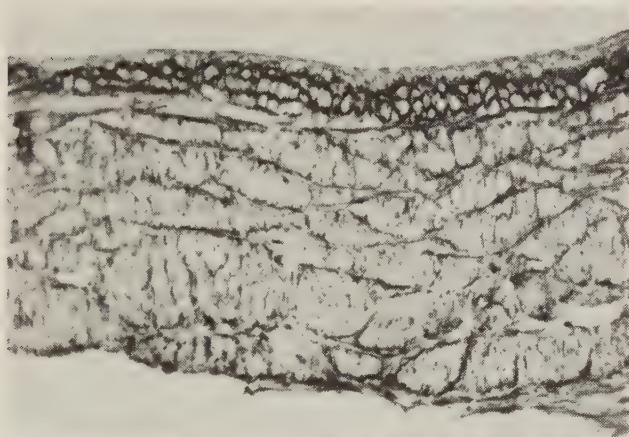


Fig. 14. Case 8. Photomicrograph of venous biopsy. Elastin. $\times 100$.

be of great value even in congenital absence of valves; indeed it may serve to keep the patient almost free of symptoms and thus fit for work.

As regards vascular surgery, division of communicating veins (*Cockett's operation*) was tried in one of the present cases and had at least a satisfactory initial effect. If phlebography reveals insufficiency of the direct communicating veins, such an operation may be of value in cases with congenital absence of valves.

The popliteal resection introduced by Bauer (1948) is not serviceable in these conditions since it is based on the premise that functioning valves exist in other venous sectors through which the blood is intended to pass. In many of the present cases valves were either entirely or partially absent even in the superficial venous system, and thus popliteal resection would have been of no avail, as evidenced by case 1, which showed no improvement after this operation.

Other types of vascular surgery, as for instance construction of valves, transplantation of veins or segments thereof, etc. may be practicable in these patients at some time in the future. Two examples of attempts along these lines may be mentioned: Röhl (1956), in experimental studies on animals, constructed competent valves by plastic surgery of the venous walls. Palma and Esperón (1960) treated femoral vein thrombosis in a few cases with good results by transplantation of the long saphenous vein from the healthy leg. The venous transplant was either anastomosed as a free graft above and below the obliteration in the external iliac and popliteal veins respectively or, in obliteration of the external iliac and/or the femoral vein, was drawn through a subcutaneous tunnel dissected above the pubis and against the deep fascia of the abdomen, after which the ends of the vein transplant were sutured in the femoral and popliteal veins respectively.

L. Conclusions

Summarizing, it may be said of the condition constituted by congenital absence of valves in the deep veins of the leg, that:

1. Hereditary predisposition may be present.
2. The condition seems to affect males and females with about the same frequency.
3. The symptoms generally have their onset around puberty.
4. The clinical picture resembles that of venous insufficiency of other etiology until ulcerations appear at the classical sites on the malleoli. As a rule varicosities are not found initially, though they may appear subsequently.
5. It is probable that in regard to the number of venous valves all variants exist, from abnormal profusion to total absence.
6. Venous insufficiency in the lower leg is a sequel of impaired venous drainage.
7. Young persons with venous insufficiency and no record of thrombosis, in whom no other explanation of the symptoms can be found, should be examined with respect to the possibility of congenital absence of venous valves.

CHAPTER 5

SUMMARY

By the term "venous insufficiency in the lower leg" the author means a chronically deficient venous drainage — of whatever etiology — in the lower leg, as well as various types of associated complications.

Approximately 2.11 per cent of the population of Sweden is estimated to have had thrombosis of the legs. Of the disability pensions granted annually in this country, 0.3 per cent currently go to patients falling in the diagnostic group comprising varicose veins and leg ulcers, defined according to the WHO nomenclature. These conditions account for the loss of a quarter of a million working days every year in Sweden. Roughly 7 per cent of the patients treated at the Dermatology Department of Karolinska Sjukhuset are suffering from leg ulcers as a result of venous insufficiency.

Venous insufficiency was long assumed to be a sequel of varicose veins; not until recent decades were post-thrombotic conditions found to be implicated at least as often as varicosities. Observations reported during the past few years have demonstrated the pathogenetic significance not only of post-thrombotic lesions of the communicating veins of the lower leg, but of non-post-thrombotic deep incompetence — for which morbid condition the term "pan-phlebotis" has also been suggested.

The normal anatomy of the venous valves in the leg is surveyed. There is no significant difference between males and females in respect to the number of venous valves; moreover, the number does not decrease progressively with mounting age.

Following a survey of previous investigations on congenital absence of valves in the deep veins of the leg, personal studies in which the author has differentiated the relevant anomaly, are reported. Twelve examples of this anomalous condition, which may be associated with an hereditary predisposition, are described. The anomaly appears to have approximately the same incidence in the two sexes. It seems probable that, as respects the number of valves, all variants exist — from abnormal profusion to total absence.

Symptoms of venous insufficiency in these cases usually have their onset around the time of puberty, probably due among other things to the growth in height occurring at that time and the consequently increased hydrostatic pressure on the tissues of the lower leg in the erect position.

The clinical symptomatology in congenital absence of venous valves largely resembles that found in venous insufficiency of other etiology. Varicose veins do not occur initially as a rule, though they may develop later. The diagnosis must be verified by phlebography.

None of the patients in this series had any history of leg injury. An incidental finding in one case was left-sided origin of the right subclavian artery; otherwise no arterial anomalies were observed. Ophthalmologic examinations revealed normal conditions and no detectable vascular anomalies in the eyegrounds. The arterial circulation of the extremities was normal in all cases. The venous in-

sufficiency in the lower legs of these patients must be attributed to impaired venous drainage. Histopathologic examination of venous biopsies from the upper surfaces of the feet in nine of these patients disclosed no lesions resembling those which, according to Bauer, are found in pan-phlebotomy.

There is no causal therapy for congenital absence of venous valves. Great value, however, attaches to consistently applied conservative therapy of the type employed in venous insufficiency of other etiology.

Résumé

Sous le terme d'«insuffisance veineuses du membre inférieur», l'auteur entend l'insuffisance du drainage veineux, de quelque étiologie qu'elle soit, ainsi que les diverses complications qui lui sont associées.

On estime que le 2,11 % de la population suédoise souffre de thromboses des veines des membres inférieurs. Dans ce pays, le 0,3 % des rentes pour incapacité de travail sont versées à des patients qui appartiennent à ce groupe d'affections comprenant notamment les varices et les ulcères de jambes (défini par la nomenclature de l'OMS). On considère en Suède que ces maladies causent chaque année la perte d'un quart de million de journées de travail.

Le 7 % environ des patients qui sont traités à la clinique dermatologique du Karolinska Sjukhuset souffrent d'ulcères de jambes résultant d'une insuffisance veineuse.

L'insuffisance veineuse a été considérée pendant longtemps comme une conséquence des varices; ce n'est que depuis quelques décades que l'on admet que les état post thrombotiques sont pour le moins aussi souvent l'origine de l'insuffisance veineuse que les varices. Les observations rapportées ces dernières années ont mis en évidence non seulement l'importance pathogénétique des lésions post thrombotiques des veines communicantes des membres inférieurs, mais aussi du facteur de l'insuffisance des veines profondes (cette dernière maladie est aussi désignée sous le terme de «pan-phlebotomy»).

L'auteur expose l'anatomie normale des valvules des veines du membre inférieur. Il ne semble pas qu'il y ait de différence notable quant au nombre des valvules veineuses dans deux sexes; en outre, leur nombre ne diminue pas avec l'âge.

L'auteur passe en revue les recherches antérieures concernant l'absence congénitale des valvules des veines profondes des membres inférieurs puis il décrit 12 cas personnels de malades présentant cette anomalie qu'il a été possible de mettre en relation avec une prédisposition héréditaire. Cette anomalie semble avoir la même fréquence dans les 2 sexes. Il semble probable que, quant au nombre des valvules, toutes les variantes peuvent exister, depuis la surabondance anormale jusqu'à l'absence totale.

En règle générale, les symptômes de l'insuffisance veineuse commencent à se manifester aux environs de la puberté. Ceci est probablement dû, entre autres, au fait que la période de croissance accélérée qui survient à ce moment, provoque une augmentation de la pression hydrostatique dans les tissus des membres inférieurs lors de la station verticale. La symptomatologie de l'absence congénitale des valvules veineuses ressemble beaucoup à celle que l'on voit dans l'insuffisance veineuse provenant d'autres étiologies.

Dans la règle, les varicosités ne se développent pas dès le début de l'affection et parfois peuvent apparaître tardivement.

Aucun des patients de cette série n'avait présenté d'accident au niveau des membres inférieurs.

Dans un cas où on a trouvé incidemment que l'artère sous clavière droite avait son origine à gauche mais aucune autre anomalie artérielle n'a été mise en évidence; l'examen ophtalmologique a montré que le fond d'œil de ces malades était normal et ne présentait aucune anomalie vasculaire.

La circulation artérielle dans les extrémités s'est révélée normale dans tous les cas. L'«insuffisance veineuse» que présentait ces patients doit être attribuée à une insuffisance du drainage des veines.

L'examen histopathologique de biopsies veineuses prélevées dans les couches superficielles de la peau des pieds chez 9 de ces patients n'a pas montré de lésions semblables à celles que Bauer avait trouvées dans la pan-phlebotomie.

S'il n'existe pas de traitement étiologique de l'absence congénitale de valvules veineuses, il faut par contre attacher une grande importance à la thérapeutique conservatrice qui est identique à celle qui est pratiquée en cas d'insuffisance veineuse d'autre origine.

Zusammenfassung

Mit der Bezeichnung »venöse Insuffizienz der unteren Extremität« meint der Autor eine chronische Störung des venösen Abflusses — gleichgültig welcher Ätiologie — der Unterschenkel sowie verschiedene Typen damit verbundener Komplikationen.

Schätzungsweise haben 2,11 % der Bevölkerung Schwedens Thrombosen der Beine durchgemacht. Von den jährlich zugesprochenen Renten wegen Arbeitsunfähigkeit entfallen 0,3 % auf Patienten, die diagnostisch nach der Definition der Weltgesundheitsorganisation in die Gruppe der varikösen Venen und Unterschenkelgeschwüre einzuordnen sind. Durch diese Krankheiten gehen in Schweden jährlich $\frac{1}{4}$ Million Arbeitstage verloren. Ungefähr 7 % der in der Hautklinik des Karolinska Sjukhuset behandelten Patienten leiden an Unterschenkelgeschwüren infolge venöser Insuffizienz.

Die venöse Insuffizienz wurde lange Zeit als Folge variköser Venen angesehen; erst in den letzten Jahrzehnten wurde bekannt, dass post-thrombotische Zustände mindestens ebenso häufig wie die Varicosis als Ursache der venösen Insuffizienz angesehen werden müssen. Beobachtungen der letzten Jahre haben nicht nur die pathogenetische Bedeutung von postthrombotischen Veränderungen der kommunizierenden Venen der Unterschenkel, sondern auch von nicht-postthrombotischen tiefen Störungen gezeigt, für die auch die Bezeichnung »Pan-Phlebotomie« vorgeschlagen wurde.

Es wird ein Überblick über die normale Anatomie der Venenklappen des Beines gegeben. Es besteht keine signifikante Differenz in der Zahl der Venenklappen zwischen Männern und Frauen; weiterhin nimmt die Zahl mit zunehmendem Alter nicht ab.

Nachdem über frühere Untersuchungen zum congenitalen Fehlen von Klappen der tiefen Beinvenen berichtet worden ist, werden die eigenen Untersuchungen mitgeteilt, in denen der Autor die wichtigen Anomalie differenziert hat. Zwölf Beispiele von anormalen Bedingungen, die mit einer hereditären Prädisposition verbunden sind, werden angeführt. Die Anomalie scheint bei beiden Geschlechtern gleich häufig zu sein. Es ist wahrscheinlich, dass hinsichtlich der Zahl der Klappen alle Varianten zwischen völligem Fehlen und abnormem Überfluss existieren.

Symptome von venöser Insuffizienz beginnen in diesen Fällen gewöhnlich um die Zeit der Pubertät, u. a. wahrscheinlich infolge des in dieser Zeit gesteigerten

Längenwachstums und des damit verbundenen erhöhten hydrostatischen Druckes auf die Gewebe der Unterschenkel bei aufrechter Körperhaltung.

Die klinische Symptomatologie bei congenitalem Fehlen von Venenklappen entspricht derjenigen der Veneninsuffizienz anderer Ätiologie. Im Beginn treten im allgemeinen keine varikösen Venen auf, jedoch können sie sich später entwickeln. Die Diagnose muss durch die Phlebographie verifiziert werden.

Keiner der Patienten bot anamnestische Hinweise für ein Beintrauma. In einem Fall wurde gleichzeitig ein linksseitiger Ursprung der rechten Arteria subclavia gefunden; im übrigen fanden sich keine Abnormitäten der Arterien.

Ophthalmologische Untersuchungen ergaben normale Verhältnisse und keine Gefässanomalien am Augenhintergrund. Die arterielle Zirkulation war in allen Fällen normal. Die Veneninsuffizienz der Unterschenkel muss auf den gestörten venösen Abfluss zurückgeführt werden. Histopathologische Untersuchungen von Venenbiopsien aus der Fussoberfläche ergaben keine Veränderungen, wie sie nach Bauer bei der Pan-Phlebois gefunden werden.

Es gibt keine ursächliche Behandlung für das congenitale Fehlen von Venenklappen. Grosser Wert kommt jedoch einer konsequent durchgeführten konservativen Therapie zu, wie sie bei Veneninsuffizienzen anderer Ätiologie angewandt wird.

Resumen

Por el término de «insuficiencia venosa de la parte inferior de la pierna» indica el autor un drenaje venoso deficiente crónico — de la etiología que sea — así como varios tipos de complicaciones asociadas.

Se calcula que un 2,11 por ciento de la población de Suecia ha tenido una trombosis de piernas. De las pensiones por incapacidad concedidas anualmente en este país, 0,3 por ciento, por lo general, van a recaer en pacientes incluidos en el grupo diagnóstico de venas varicosas y úlceras de piernas, definido de acuerdo con la nomenclatura de la WHO. Estas afecciones suponen la pérdida de un cuarto de millón de días de trabajo al año en Suecia. Aproximadamente, el 7 por ciento de los pacientes tratados en el Departamento de Dermatología del Karolinska Sjukhuset padecen úlceras de pierna como consecuencia de insuficiencia venosa.

La insuficiencia venosa fué atribuída hace tiempo a una secuela de venas varicosas; y hasta las últimas décadas no se vió que eran afecciones post trombóticas por los menos con igual frecuencia que las varicosidades. Observaciones hechas durante los últimos años han demostrado la importancia patogénica no sólo de las lesiones post-trombóticas de las venas comunicantes de la parte inferior de la pierna, si no también de insuficiencias profundas no post-trombóticas, para cuyo cuadro mórbido se ha sugerido también el término de «pan-flebois».

Se examina la anatomía normal de las válvulas venosas de la pierna. No existen diferencias notables entre hombres y mujeres respecto al número de válvulas venosas; además, el número no disminuye de modo progresivo a medida que la edad aumenta.

Después de una revisión de las investigaciones previas sobre la ausencia congénita de válvulas en las venas profundas de la pierna, el autor expone sus estudios personales en los que ha distinguido esta anomalía. Doce ejemplos, describe, en los que esta anómala situación puede asociarse con una predisposición hereditaria. La anomalía parece ser aproximadamente de la misma frecuencia en los dos sexos. Parece probable, en lo que al número de válvulas se refiere, que

existan toda clase de variantes desde una abundancia anormal hasta la total ausencia.

Los síntomas de insuficiencia venosa en estos casos suelen comenzar a la época de la pubertad, probablemente debido entre otras razones al aumento de la talla que suele tener lugar en esta edad, y al consiguiente aumento de presión hidrostática de los tejidos de la parte inferior de la pierna con la estancia en pié.

La sintomatología clínica en la ausencia congénita de válvulas venosas semeja grandemente a la de la insuficiencia venosa de otras etiologías. Las venas varicosas no es de regla que aparezcan al comienzo, pueden formarse más tardíamente. El diagnóstico debe hacerse por flebografía.

Ninguno de los enfermos de esta serie había tenido historia de herida en pierna. Un hallazgo fortuito en uno de nuestros casos fué el origen en lado izquierdo de la arteria subclavia derecha; fuera de esto no se observó anomalía arterial alguna. Los exámenes optalmológicos revelaron normalidad y no descubrieron anomalías vasculares en el fondo de ojo. La circulación arterial en las extremidades fué normal en todos los casos. La insuficiencia venosa en parte inferior de piernas de estos pacientes debe ser atribuída a drenaje defectuoso en venas. El examen histopatológico de las biopsias venosas de caras superiores de piés en nueve de estos pacientes no mostraron lesiones análogas a las que según Bauer se encuentran en la panflebosis.

La ausencia congénita de válvulas venosas no tiene terapéutica causal. Sin embargo se concede gran valor a la terapéutica conservadora oportuna del tipo de la empleada en la insuficiencia venosa de otra etiología.

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PART II

ROENTGENOLOGIC INVESTIGATIONS

CHAPTER 1

BRIEF BACKGROUND

In an earlier paper (1959) presented in collaboration with H. Gentile, we reported total congenital absence of venous valves in the arms and legs of two sisters. Ten similar cases have since been collected. The aim of this paper is to report the phlebographic findings in all twelve cases and to discuss the serviceability of various phlebographic methods of establishing the presence of valves in the veins.

The normal anatomy of the venous valves of the leg has been described in Part I (Lodin), which also reviews previous investigations concerning congenital absence of venous valves. Aside from Lodin *et al.* (1959), only Hodgkin and Cooper (1836) have previously described total absence of valves in the venous system; other authors dealing with this anomaly have been concerned merely with partial absence of valves. Only a few of these authors have used phlebography in their investigations. Luke (1941, 1951), for instance, reported four cases of what he termed "a new disease entity", associated with absence of valves in the deep veins of the legs distal to the opening of the long saphenous vein. Neither his phlebograms nor his text indicate, however, whether these cases were in fact totally avalvular or whether they constituted variants with a small number of valves. It is not impossible that they were cases of panphlebotomy (Bauer, 1960) or of post-thrombotic conditions, although the roentgenograms militate against such a possibility. The examinations had been performed in one case with the patient recumbent and contrast injection via direct puncture of the femoral vein, and in three cases with the patient erect, during Valsalva's maneuver, the injection being given via a catheter inserted into the long saphenous vein.

Norman (1956) reported four cases with total absence of valves in the deep veins of the legs proximal to the opening of the short saphenous vein. He surmised that the avalvular condition was congenital. The diagnosis had been established with the aid of ascending phlebography with the patient recumbent.

May and Nissl (1959) in their monograph on phlebography of the legs, showed a roentgenogram from a case with congenital absence of valves in the femoral and popliteal veins. The examination method they used was ascending phlebography with the patient semi-erect. They referred the reader to Luke (1941) and added only that such anomalies may be found. In the same book, in the chapter on retrograde phlebography during straining, Martinet stated that of 222 patients examined, nine exhibited incompetence of the deep veins due to absence of valves. He called attention, however, to the difficulties of evaluation and said he had been unable to ascertain whether the absence of valves was congenital or post-thrombotic.

Moore (1951) found absence of valves in the deep veins of 57 of 84 examined legs with no history of thrombosis. He had used ascending phlebography with the patient recumbent, in accordance with a technique which he had described in 1949. He concluded that the condition was post-thrombotic.

Warwick (1930) conducted phlebographic examinations of cadavers and demonstrated partial absence of valves in the deep veins of the lower leg, particularly the posterior tibials.

Some authors have discussed the possibility of demonstrating by phlebographic methods the presence of valves in the veins, the number of valves in different venous sectors, and valvular incompetence. They have used in their investigations both ascending and retrograde phlebography. Ascending phlebography has been carried out not only in recumbent patients with or without Valsalva's maneuver (Moore, 1949; Gryspeerdt, 1953), but in seated (Lindblom, 1941; Höjensgård, 1949) and in erect subjects (Greitz, 1954; May and Nissl, 1959). Retrograde phlebography with injection of contrast medium directly or indirectly into the femoral vein during proximal compression of the vein or during Valsalva's maneuver, has been done both in recumbency (Luke, 1941; Bauer, 1948; Sandegård, 1949; Gullmo, 1956) and with the patient either semi- or fully erect (Luke, 1951; Sylwan, 1951; Ödman, 1952; Lodin *et al.*, 1959; Martinet, 1959). The valves have also been studied by means of tilt phlebography (Dohn, 1958; Mathiesen, 1958 b).

Of the authors mentioned above, only a few have discussed the absence of venous valves. Greitz (1955 a) for example, observed no valves in the femoral vein in some of his cases, but attributed this to shortcomings in the phlebographic technique. Mathiesen (1958 a) examined 470 patients by tilt phlebography and observed that "in no case did there seem to be sufficient evidence for diagnosing an idiopathic absence of valves". Thus, in the literature little attention has been devoted to the congenital absence of venous valves and no detailed reports of the phlebographic findings have been published.

CHAPTER 2

MATERIAL AND METHODS

The series consists of twelve subjects — five females and seven males. All of them were examined during the period 1958—1960 at the central roentgen diagnostic department of Karolinska Sjukhuset. The age at initial examination ranged from 9 to 41 in the females and from 12 to 37 in the males. Further details of the series will be found in Part I.

The examinations comprised both legs in all cases. In one patient, however, one of the lower legs was not included, and in another the veins were not studied proximal to the middle third of the thighs. Thus 24 legs, including the pelvic veins up to the inferior vena cava, were examined — 21 of them completely.

Table 1. *Roentgenologic examinations of the veins.*

Case	Ascending phlebography of legs		Retrograde phlebography of legs		Pelvic phlebography	Cavography
	Right	Left	Right	Left		
1	+	+	+	+	+	+
2	+	+	+	+	+	
3	+	+	+	+	+	
4	+	+	+	+	+	
5	+	+	+	+	+	
6	+	+				
7	+		+	+	+	+
8	+	+	+	+	+	
9	+	+	+	+	+	
10	+	+	+	+	+	
11	+	+	+	+	+	+
12	+	+	+	+		

The following examinations were undertaken: Ascending phlebography of the legs, pelvic phlebography, retrograde phlebography of the legs, and cavography (see table 1). In addition, nine of the patients were subjected to phlebography of the arms.

A. Ascending Phlebography of the Leg

(Figures 1, 2, 3.)

For this examination the subject was seated with his lower legs vertical, in accordance with the method described by Lindblom (1941). The radiopaque

substance, 40 cc of 45 per cent Urografin® (a triple-iodine contrast medium of diatrizoate type), was injected rapidly into a superficial vein in the distal part of the dorsum pedis, usually the great toe. Five phlebograms were taken, the first of them immediately after the injection and the others at intervals of approximately 10—15 seconds. The initial phlebogram comprised the ankle and lower leg, and the remainder the lower leg, knee and distal half of the thigh. The subject remained seated without moving his leg until the third phlebogram had been taken. Prior to and during the taking of the third phlebogram, he performed Valsalva's maneuver. Before the fourth phlebogram he contracted his calf muscles a few times. This procedure was repeated, and was followed by Valsalva's maneuver, for the fifth phlebogram.

B. Pelvic Phlebography

(Figures 4 and 5.)

This examination was performed with the subject recumbent, though, in one case, both in the erect and the recumbent position. The contrast medium, totalling 50 cc of 30 per cent Urografin®, was injected rapidly and simultaneously into both external iliac veins through catheters introduced percutaneously via the femoral veins *ad modum* Seldinger (1953). Four phlebograms comprising the pelvic and hip region, and including the femoral vein down to a point just distal to the opening of the deep femoral vein, were then taken. When approximately 10 cc of the contrast medium had been injected the subject began to perform Valsalva's maneuver, and continued it throughout the examination. Six patients also underwent pelvic phlebography by the method of Helander and Lindbom (1959 a), i. e.; with compression of the inferior vena cava instead of Valsalva's maneuver. In four of these cases, however, the injection was given manually.

C. Retrograde Phlebography of the Leg

(Figure 6.)

This examination was performed on the same occasion as pelvic phlebography, with the patient erect in a Forsell stand. The contrast medium, 20—30 cc of 60 per cent Urografin®, was injected slowly into the external iliac vein through the catheter inserted percutaneously via the femoral vein. When half the dose of contrast had been injected, the subject began to perform Valsalva's maneuver and continued it throughout the examination. The first phlebogram, comprising the hip region and proximal part of the thigh, was taken immediately after conclusion of the injection. The second, which comprised the distal part of the thigh, the knee and proximal portion of the lower leg, and the third which comprised the lower leg alone, were taken as soon as possible after the first. In a few instances further phlebograms were taken of the lower leg. Fluoroscopic screening provided guidance for positioning before each exposure. Eight of the patients underwent, additionally, retrograde phlebography of the legs while recumbent.

D. Cavography

For this examination the method reported by Helander and Lindbom (1959 b) was employed, though in one case the contrast medium was injected manually.

E. Ascending and Retrograde Phlebography of the Arm

In these examinations the patients stood in a Forsell stand, their arms hanging. The contrast medium consisted of 20 cc of 30 per cent, 45 per cent, or 60 per cent Urografin®. In the retrograde examinations the agent was injected slowly into the axillary vein through a catheter inserted via the basilic vein *ad modum* Seldinger (1953). Valsalva's maneuver was performed throughout the examination, beginning immediately after the start of the contrast injection. In the ascending phlebographies the contrast medium was injected fairly rapidly into a superficial vein of the hand or wrist. In these cases Valsalva's maneuver was initiated immediately after the contrast injection had terminated. Phlebograms were taken of the shoulder and upper arm region, the elbow region, and the forearm and hand.

Ascending phlebography of the legs was bilateral in all except one subject, who allowed only one leg to be examined. Retrograde phlebography of the legs and pelvic phlebography were undertaken in all subjects with the exception of a nine-year-old girl, who was excluded because of her age. Three patients underwent cavography and nine phlebography of the arms, in five of them bilateral.

CHAPTER 3

RESULTS

It was observed at the ascending phlebographies that the contrast medium passed in the normal way into the deep and superficial veins, and also entered some muscle veins and deep communicating veins (figs. 1 and 2). The contrast passage was in most cases normal, as was disappearance from the veins when the subjects had contracted their calf muscles a few times. In occasional cases with superficial varicose veins, passage of the contrast medium was slightly retarded. On Valsalva's maneuver, the contrast was forced retrogradely from the femoral vein down into the lower leg. As a result the femoral vein in some cases was almost emptied of radiopaque substance while e. g. the superficial and muscle veins showed increased contrast filling (fig. 3).

In pelvic phlebography and retrograde phlebography of the legs the contrast was rapidly forced retrogradely when the subject performed Valsalva's maneuver. The longer the patient was able to continue this maneuver, the more did the retrograde contrast filling increase. Indeed, in some cases the contrast reached as far as the ankle (fig. 6). All variants were observed, ranging from complete retrograde filling of deep and superficial veins, of muscle veins in the gluteal region, in the thighs and lower legs, as well as almost total filling in the region of distribution of the internal iliac vein (fig. 4), to filling only of the deep venous trunks, i. e., the femoral, popliteal, and more or less of the lower leg veins. The pelvic phlebographies with Valsalva's maneuver and those with vena cava compression *ad modum* Helander and Lindbom, differed substantially in regard to the distribution of contrast, with the exception of one case in which the results with the two methods were practically identical. The Helander-Lindbom technique was associated with opacification of the internal iliac, sacral, vertebral and other veins, but no contrast filling (except for one case with very slight filling) of the external iliac and femoral veins (fig. 5 a). When the same patients were examined without vena cava compression but during Valsalva's maneuver, the internal iliac vein and its branches showed only very slight filling, whereas there was complete opacification of the external iliac and femoral veins and the veins of the lower leg, and in some cases, of gluteal muscle veins as well (fig. 5 b). Practically all cases in which retrograde phlebography of the legs was done with the subjects both erect and recumbent, showed better retrograde contrast filling in the former position. In the other cases the results were equivalent.

The incidence of venous valves is shown in table 2. In case 4 the right popliteal vein was duplicate. One of the duplicate veins contained a valve; the other was avalvular, and the likewise valveless, dilated short saphenous vein opened into it. In case 6 only the distal two-thirds of the thighs were examined.

Table 2. Incidence of valves in the different veins.

Case	Common iliac		External iliac		Femoral		Deep femoral		Popliteal		Deep veins of lower leg		Long saphenous		Short saphenous	
	Right	Left	Right	Left	Right	Left	Right	Left	Right	Left	Right	Left	Right	Left	Right	Left
I	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
2	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
3	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
4	0	0	0	0	0	0	n	n	n0	0	oc	af	0	d	d	0
5	0	0	0	0	0	0	0	0	0	0	oc	oc	un	un	?n	?n
6	—	—	—	—	0	0	—	—	0	0	0	0	0	0	oc	0
7	0	0	0	0	0	0	n	n	0	—	oc	—	n	d	d	—
8	0	0	0	0	0	0	n	n	0	0	af	af	—	—	un	un
9	0	0	0	0	0	0	n	n	0	0	af	af	d	d	d	d
10	0	0	0	0	0	0	oc	n	0	0	af	oc	0	0	0	0
11	0	0	0	0	0	0	af	af	0	0	0	0	0	0	0	0
12	0	0	0	0	0	0	oc	0	0	0	oc	oc	un	un	0	d

0 = avascular

oc = an occasional valve

af = abnormally few valves

— = not examined

un = unevaluable

d = dilated and not reliably evaluable

n = normal number of valves

Case 7 exhibited an incomplete valve, probably with only one leaflet, in the right femoral vein; the left leg had a normal incidence of valves. The deep veins of thirteen lower legs were valved, the incidence varying from an occasional valve or so for the entire lower leg to between one and four in the different venous trunks.

In no case did retrograde phlebography reveal any valves which had not been detected at ascending phlebography.

The inferior vena cava was normal in the three cases in which it was examined.

The deep veins were of normal caliber in all subjects except one whose posterior tibial veins were slightly dilated in one leg, though not more than e. g. Greitz (1955 a) considered to be within the normal range. In most cases the deep veins of the lower leg had a strikingly thin appearance, due to absence of valvular sinuses (fig. 1).

The variations in valvular contour ranged from the conspicuous pouches of normal valves to the type of valve with no bulging sinus but only leaflets in the vascular lumen. Since in the retrograde phlebographies the contrast medium was arrested at the first functioning valve, it was not always possible to ascertain if valves of the second-named type functioned normally. Some of them appeared to be competent but others were visualized so unsatisfactorily that insufficiency was suspected. No further examinations were undertaken, however, to confirm this suspicion.

The subjects exhibited no pathologic changes such as are reportedly associated with pan-phlebotomy. Nor were any lesions suggestive of post-thrombotic conditions observed.

Four of the phlebographically examined arms (three subjects) were devoid of valves in both the deep and the superficial veins, and the contrast medium, injected into the axillary vein, passed retrogradely as far as the hand. One arm showed only a single valve or so in the deep veins, the superficial veins being avalvular. Two arms had occasional valves in both the deep and the superficial veins, while in seven arms the incidence of valves was probably normal.

Thus two of the twelve phlebographed subjects were devoid of valves in all veins examined, both in the arms and legs, while one had a solitary valve or so in the deep femoral vein and likewise in the deep veins of one arm. One patient was totally avalvular in the legs but had occasional valves in the deep and superficial veins of the arms. In one case there was a normal incidence of valves in the femoral and popliteal veins of one leg (lower leg not examined); whereas the contralateral femoral vein had one incomplete valve, the popliteal vein was avalvular, and the veins of the lower leg showed only a single valve or so. (Cf. table 2.)

DISCUSSION

There are, as will be seen from the review of the literature, a great many methods for roentgen examination of the veins of the leg. Lindblom (1941) showed that ascending phlebography should be performed with the leg vertical, a finding that was further emphasized by Kjellberg (1943), Höjensgård (1949), and Greitz (1954), among others. Mainly due to the major difference in specific gravity, contrast media and blood do not mix without turbulent flow. Optimal admixture is obtained with the leg in the vertical position (Kjellberg, 1943). Because of the high specific gravity of the contrast media these are able to flow retrogradely past incompletely closed valves. Hence retrograde contrast filling of venous areas does not necessarily connote absence or incompetence of valves — a consideration which has been emphasized by Kjellberg (1943), Luke (1943), Sandegård (1949) and Greitz (1955 b), among others. Only by visualization of the valves, therefore, is it possible to estimate their presence.

Højensgård (1949) pointed out that if valves are to be phlebographically demonstrable, a vertical position of the leg is required. The veins contain a greater volume of blood in the vertical than in the horizontal position, and moreover the venous circulation is slower. Hence the valves are partially closed, except when the circulation is accelerated by muscle contractions. The possibilities of valve visualization will be further enhanced if the subject performs Valsalva's maneuver during ascending phlebography, for the retrograde movement of blood closes the valves (Gryspeerd, 1953). The layering tendency of contrast medium contributes to visualization of the valves under these conditions (fig. 3 c).

The ascending phlebographies in this investigation were carried out with the lower legs vertical. Opinions vary as to the most suitable injection site in such examinations. We elected to give the injections in a superficial vein on the dorsum pedis; others such as Arnoldi and Bauer (1960), however, prefer to inject the contrast medium into the spongiosa of the calcaneus or the lateral malleolus. The fact that the first phlebograms were taken with the legs relaxed and without contraction of the calf muscles, and the third phlebogram during Valsalva's maneuver, facilitated demonstration of valves. Under these circumstances the venous circulation was either retarded or arrested and the valves were fully or partially closed, the conditions thus being conducive to layering of the contrast medium in the valve pouches (fig. 3). The valves would probably have emerged even more distinctly with the use of tilt phlebography, but in this investigation no tilt table was available. A drawback of tilt phlebography is the frequent absence of contrast filling of the anterior tibial veins. There is no reason to assume that the present method is less reliable than tilt

phlebography for demonstration of valves. The competence of the existing valves, however, could not be evaluated, nor in all cases could the sufficiency of the deep communicating veins.

Ascending phlebography is not a dependable method for detection of venous valves in the thigh and pelvis. Greitz (1955 a) in his studies of its potentialities, found that it did not invariably permit visualization of valves in the femoral vein. He attributed this shortcoming to deficiencies of the technique employed. The contrast medium is injected in the foot, and the higher it ascends the more it is diluted by admixture with blood. The velocity of flow is greater in the proximal parts of the leg, and hence the valves are more open — a fact which makes visualization even more difficult. Gryspeerdt (1953) performed his examinations with the patients recumbent during Valsalva's maneuver. In his opinion the straining caused a rapid retrograde flow of the blood, conducive to mixture with the contrast medium. This insured satisfactory opacification and thus facilitated detection of venous valves despite the recumbent position. The avalvular cases reported by Norman (1956) had been examined by this method.

It is evident from the foregoing that the best methods for visualization of venous valves in the pelvis and thigh are pelvic phlebography and retrograde phlebography of the leg, if done during performance of Valsalva's maneuver and, as respects the thigh, with the subject erect. In the available literature on pelvic phlebography we have found no mention of its serviceability for demonstration of valves in the common and external iliac veins or in the femoral vein. The method best suited for evaluation of the pelvic veins is that reported by Helander and Lindbom (1959 a). The present studies were conducted with a modification of that method, the blood flow through the inferior vena cava being arrested by means of Valsalva's maneuver instead of by vena cava compression. The advantage of the modified method is that Valsalva's maneuver checks the flow even in the sacral, presacral and vertebral veins as well as in the azygos vein. Helander and Lindbom in their first investigations used the method described here, but subsequently adopted compression of the vena cava and injection of the contrast medium with Gidlund's pressure syringe. Filling of the internal iliac, sacral and vertebral veins was better with the latter method, though the difference was not very great. The satisfactory filling of those veins was, in their opinion, due to the fact that the contrast injection increased the venous pressure to a point where the valves closed in the proximal part of the femoral vein and the radiopaque substance was forced into the internal iliac and the sacral veins. They generally observed no filling of the external iliac vein. In the present series, comprising patients with absence of valves in the external iliac and the deep veins of the legs, the two methods of pelvic phlebography differed substantially in all except one subject, a twelve-year-old boy who was unable to perform Valsalva's maneuver correctly. The remaining subjects showed no filling of the external iliac and femoral veins on compression of the vena cava, whereas the other pelvic veins were well filled. The reverse was the case, however, when the examination was carried out during Valsalva's maneuver (fig. 5), for then the blood flow was checked even in the vertebral veins and the azygos vein. When no valves existed in the common and external iliac veins or the femoral vein, the contrast medium and blood were forced

along the path of least resistance, i. e., into the leg. Valsalva's maneuver, furthermore, produced a retrograde movement of blood in the inferior vena cava and not merely an arrest of the venous return flow. The investigations demonstrated too, that the method of Helander and Lindbom (1959 a) gives filling of the internal iliac and the vertebral veins not because of valve closure in the femoral vein, but simply because the compression has checked the flow in the inferior vena cava and hence the blood must take the only remaining route, i. e., the above-mentioned veins. With the modified examination used here, a rapid elevation of pressure in the pelvic veins is not desirable. The aim is by means of straining to bring about a retrograde movement of blood and contrast medium, serving to close any venous valves that may be present. It is therefore more advantageous to inject the radiopaque substance by hand than by a pressure syringe.

The same considerations that apply to demonstration of venous valves in ascending phlebography of the legs, are also valid for retrograde phlebography with the patient in the erect position and performing Valsalva's maneuver (fig. 6). An advantage of the method employed here is that it can be carried out on the same occasion as pelvic phlebography. The fact that the injection is given through a catheter instead of through a needle facilitates positioning of the subject. Since the injection itself is antegrade and the contrast medium is forced retrogradely by the straining in Valsalva's maneuver, the spread of contrast into all venous areas is more satisfactory than that following retrograde injection as performed by e. g. Luke (1943), Bauer (1948), Sylwan (1951) and Ödman (1952). In retrograde injection by the methods described, the femoral vein is punctured distal to the inguinal ligament. Hence neither the proximal parts of the femoral vein nor the iliac veins are examined. A similar situation pertains to those methods in which the contrast medium is injected retrogradely through a catheter inserted via the long saphenous vein. Examination of the patient in the vertical instead of the horizontal position has the advantage that the contrast medium, due to its high specific gravity, facilitates retrograde passage. This is quite evident from the fact that with this procedure the contrast medium can be made to pass retrogradely even if normally functioning valves are present. For this reason the vertical position is unsuitable for evaluation of mild degrees of valvular incompetence. According to Gullmo (1956) it is advantageous to have the patient recumbent, since in the erect position the leg veins contain so much blood that only a small additional quantity can be forced retrogradely into the leg. However, both the present examinations and many others on patients with a normal incidence of venous valves — who were examined in both positions — indicate that retrograde contrast filling is better, or at least as satisfactory, in vertical position. This may be due partly to the fact that Valsalva's maneuver can be performed more effectively while standing, and partly to the fact that the high specific gravity of the contrast medium facilitates retrograde passage. The volume of the veins evidently may be so appreciable that even in the erect position a substantial additional amount of blood can be forced into the legs. Because of technical considerations, however, phlebograms are often of better quality in the horizontal than in the vertical position. One disadvantage of examination with the patient standing is that it is not practicable with the leg relaxed; if it were, the

retrograde examination would be further facilitated. The difference between horizontal and vertical examinations is particularly marked in subjects who are unable to perform Valsalva's maneuver correctly.

The subjects in this series, as is evident from the foregoing, were so examined that venous valves, if present, must have been fully or partially closed. Since the legs, moreover, were examined in the vertical position and since the relevant contrast media tend to form layers in the vasal lumina, the possibilities of demonstrating valves were excellent. (*Cf.* figs. 1, 2 and 3). The relevant methods appear, therefore, to be suitable for visualization of normally developed valves, whether competent or incompetent.

The incidence of valves can be determined with considerable assurance in those large venous trunks, proximal to and including the popliteal vein, which are best evaluated by means of retrograde phlebography. This observation is fully consonant with results obtained in normals.

In retrograde examinations the phlebograms of the lower legs were not always of satisfactory quality, due partly to technical factors and partly to the subjects' inability to maintain Valsalva's maneuver for a sufficient length of time. Evaluation of the lower legs with respect to venous valves was, therefore, based largely on ascending phlebography. At this examination even the superficial veins were contrast-filled to a large extent. Hence it is not impossible that occasional valves may have escaped detection should the pertinent venous area have been concealed either by an opacified superficial vein, or by the compact border of the tibia or fibula. Since, however, each of the subjects examined had less than one-half the number of valves hitherto regarded as the lower limit of normal, their classification as cases of total or subtotal congenital absence of venous valves should be fully reliable.

CHAPTER 5

CONCLUSIONS

For elucidation of the incidence of venous valves in the leg, examination with the leg vertical while the subject performs Valsalva's maneuver is the best method. It is advisable to employ a combination of ascending and retrograde phlebographies. With the use of the methods described here, it should be possible to establish the incidence of valves with considerable accuracy.

From the series reported, it is evident that (1) congenital absence of venous valves in the leg is not altogether an uncommon condition*; (2) all variants of veins probably exist, from those with an excess of valves to those completely devoid of valves; and (3) the absence of valves may be limited to superficial and/or deep veins in only one or in both legs.

* Of the phlebographies performed at the roentgen diagnostic department of Karolinska Sjukhuset during the period 1958—1960, about 1 per cent showed congenital avaluular conditions.

CHAPTER 6

SUMMARY

A series of 12 patients with total or subtotal congenital absence of valves in the deep veins of the leg is reported. The serviceability of various phlebographic methods for demonstration of venous valves is discussed. It is concluded that phlebographic examinations, whether ascending or retrograde, should be done with the leg in the vertical position and with the aid of Valsalva's maneuver.

Résumé

L'auteur rapporte les cas de 12 patients présentant une absence totale ou subtotale des valvules des veines profondes des membres inférieurs, d'origine congénitale.

Il discute l'utilité pratique des différentes méthodes de phlébographies pour l'examen des valvules veineuses. Il en conclut que les phlébographies, tant ascendantes que rétrogrades, doivent être effectuées, les jambes étant en position verticale et avec l'aide de la manœuvre de Valsalva.

Zusammenfassung

Es wird über eine Serie von 12 Patienten mit totalem oder subtotalem, angeborenen Fehlen von Klappen in den tiefen Beinvenen berichtet. Die Brauchbarkeit von verschiedenen phlebographischen Methoden für die Demonstration von Venenklappen wird diskutiert. Es wird gefolgert, dass phlebographische Untersuchungen, ob ascendierend oder retrograd, in vertikaler Stellung des Beines sowie unter Zuhilfenahme des Valsalva'schen Versuches durchgeführt werden sollten.

Resumen

Se describe una serie de 12 pacientes con ausencia congénita total o subtotal de válvulas en las venas profundas de la pierna. Se discute la utilidad de varios métodos flebográficos para la demostración de válvulas venosas. Se llega a la conclusión de que los exámenes flebográficos, ascendentes o retrógrados, deben ser practicados con la pierna en posición vertical y con el auxilio de la maniobra de Valsalva.

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FIGURES

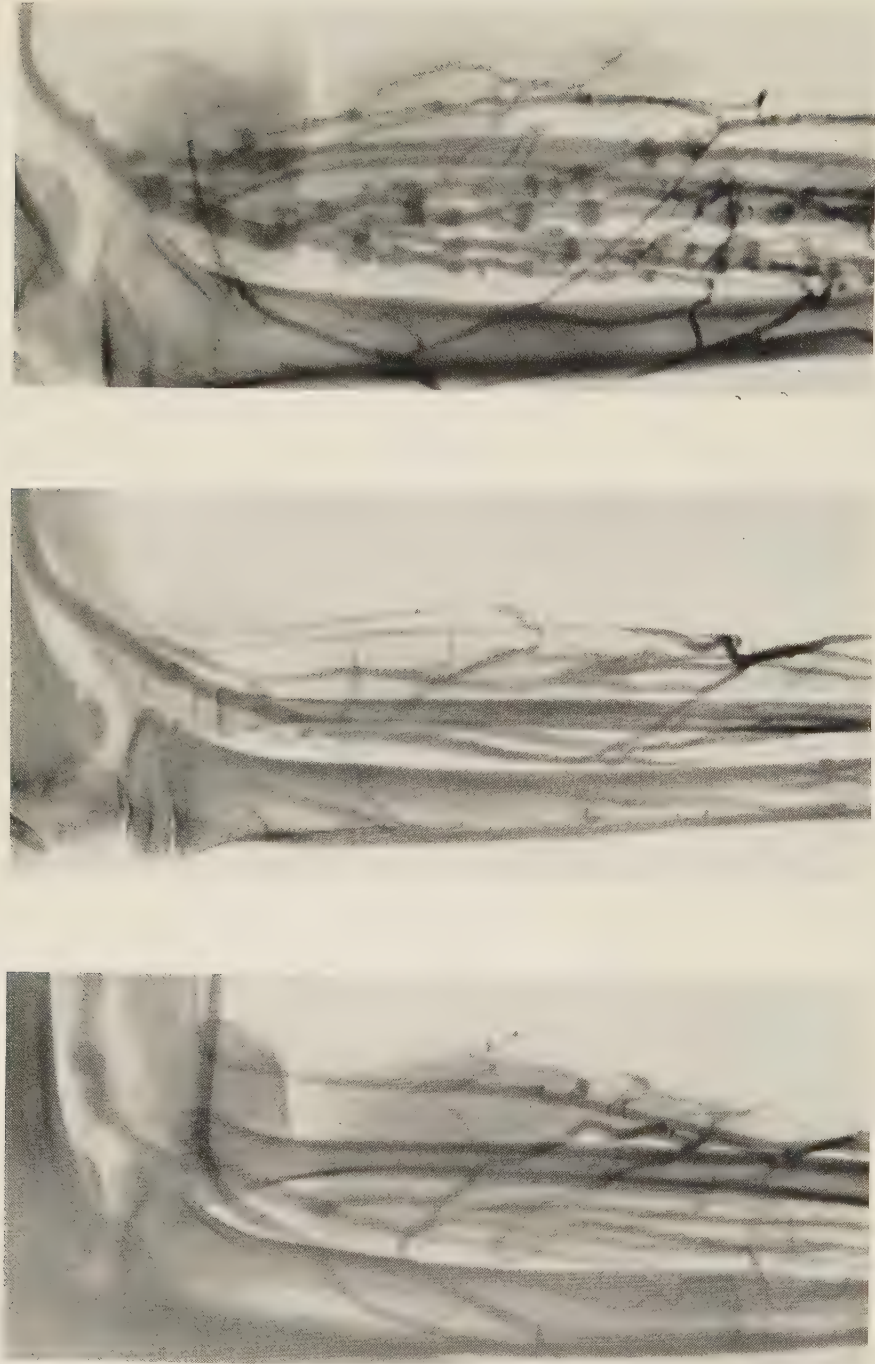


Fig. 1 c

Fig. 1 b

Fig. 1 a

Fig. 1. Ascending phlebography. (a) Case 1. There are no valves either in the deep or the superficial veins. The popliteal area shows collaterals following venous resection. (b) Case 6. The deep veins contain no valves; the superficial veins an occasional valve or so. (c) Phlebogram of a normal, showing abundance of valves and conspicuous valvular sinuses.

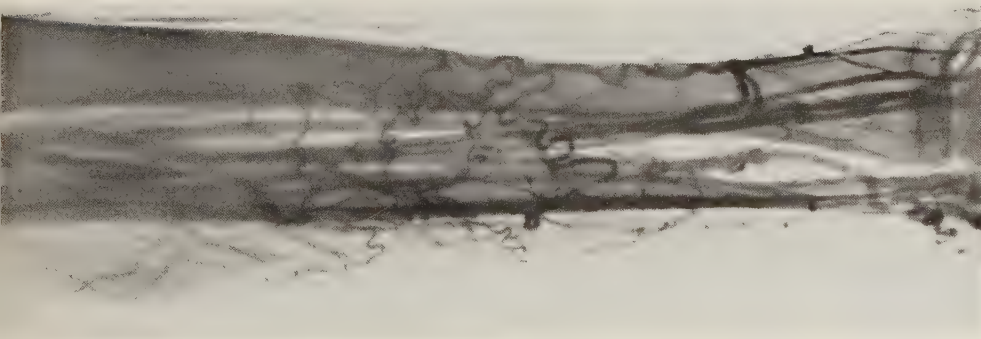


Fig. 2 a

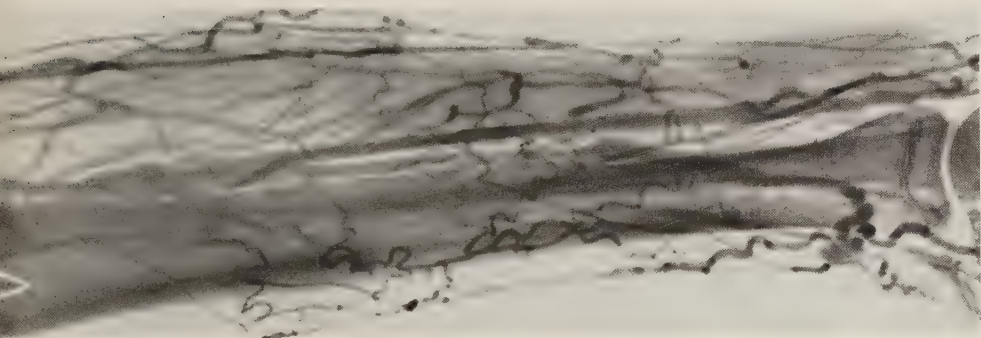


Fig. 2 b

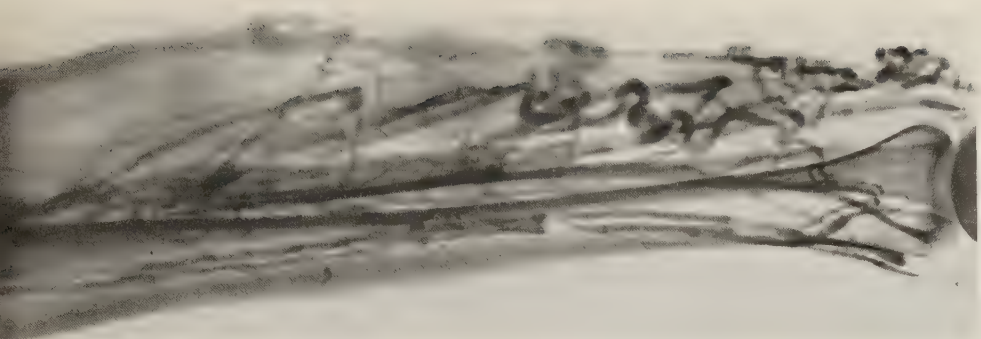


Fig. 2 c

Fig. 2. Ascending phlebography. (a) Case 11. Antero-posterior view of lower leg, showing no valves either in the deep or the superficial veins. (b) Lateral view of lower leg in the same case. (c) Phlebogram of patient with normal incidence of valves in the deep veins of the lower leg. No valves were discernible in the popliteal and femoral veins. (See Part I, page 50.)

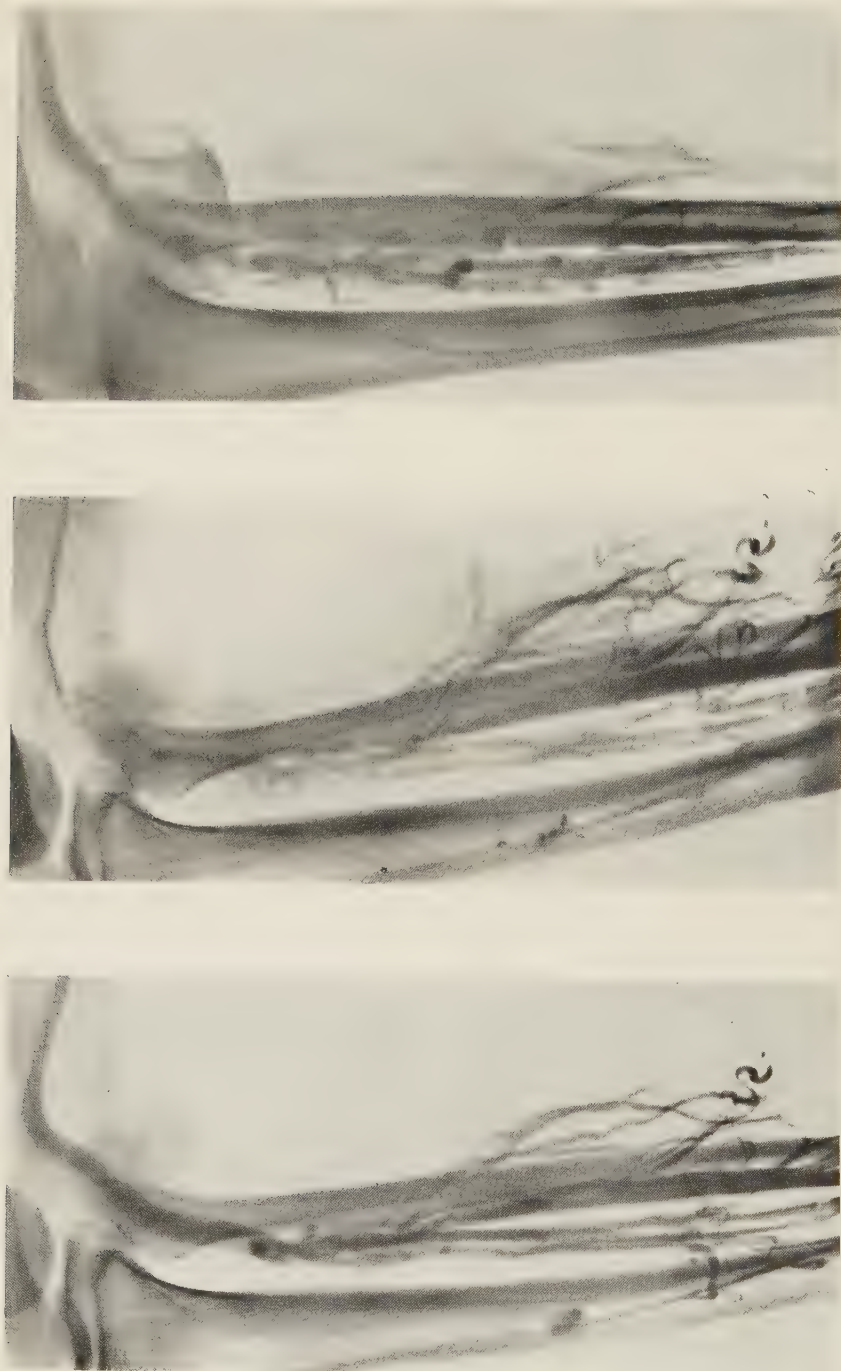


Fig. 3 a

Fig. 3 b

Fig. 3 c

Fig. 3. Ascending phlebography. (a) Case 10. A single valve or so can be observed in the deep veins of the lower leg. The phlebogram was taken prior to Valsalva's maneuver and before contraction of the calf muscles. (b) Phlebogram taken during Valsalva's maneuver at the same examination. Femoral and popliteal veins as well as proximal part of the lower-leg veins have been cleared of contrast, but there is increased filling of muscle veins and superficial veins. (c) Phlebogram of a normal, taken during Valsalva's maneuver. The contrast medium is not forced retrogradely. The valves are visualized distinctly in the popliteal vein and veins of the lower leg.

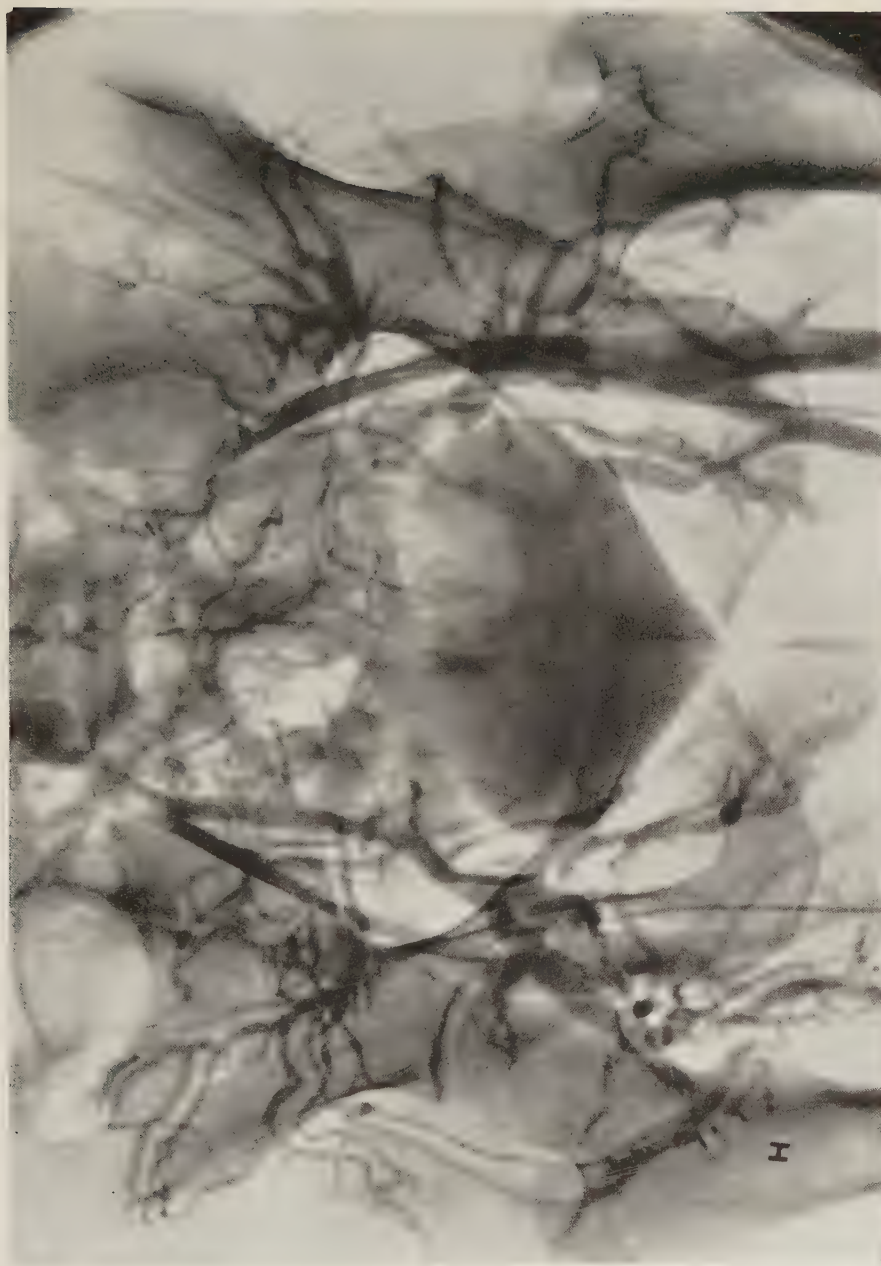


Fig. 4. Pelvic phlebography in the erect position. Case 1. Retrograde filling of both deep and superficial veins of the leg, gluteal muscle veins, femoral circumflex veins, and internal iliac veins and their branches.

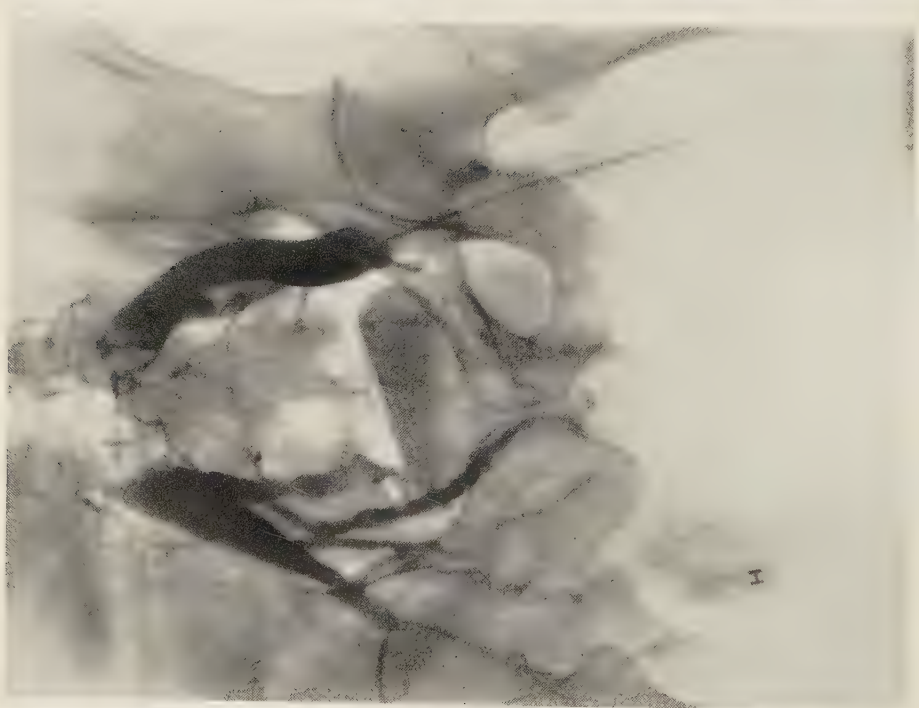


Fig. 5 a

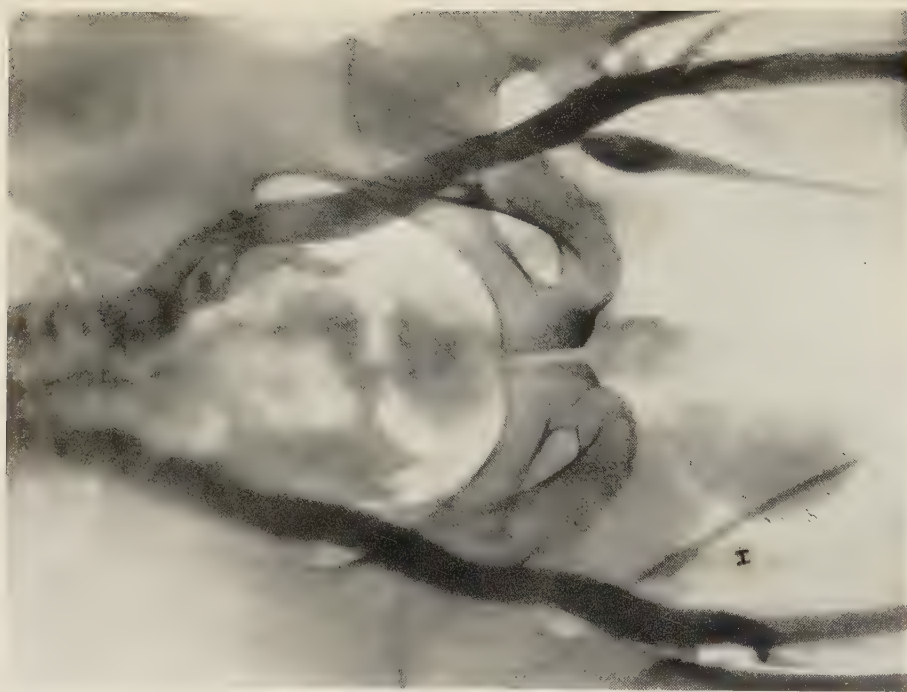


Fig. 5 b

Fig. 5. Pelvic phlebography. Case 4. (a) Examination *ad modum* Helander and Lindbom. There is no filling of the femoral veins, and the contrast passes via the internal iliac and sacral veins to the vertebral veins. (b) Examination during Valsalva's maneuver (without compression of the inferior vena cava). Very slight filling of the internal iliac veins; retrograde filling of the femoral veins and their branches and of the long saphenous veins.

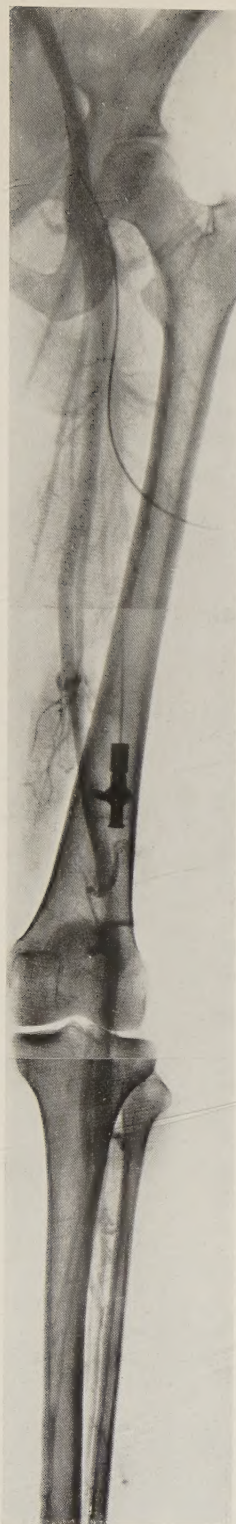


Fig. 6. Retrograde phlebography in the erect position. Case 2. The contrast medium has been injected into the external iliac just distal to the opening of the internal iliac vein; on Valsalva's maneuver, it is forced retrogradely as far as the foot. No valves are discernible. The long and short saphenous, deep femoral, muscle veins, etc. are well visualized.

